

'BAMBINO'

Statuette in bronze by Vittorio Ghiberti (fifteenth century)
From a private collection in Florence (*photograph by Ian T T Higgins*)

THE UROLOGY OF CHILDHOOD

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FOREWORD

THE past twenty five years have seen a very considerable advance in children s urology

Improved instruments of suitable size increasing skill in their use technical advances in radiology bacteriology biochemistry and pathology have combined to enable us to investigate and understand the urological disorders of childhood with an accuracy denied to our predecessors The control of infection both of the inter-current infections in hospital environment and of those of the urinary tract itself together with the increased facilities available for the restoration and main tenance of vitality in sick children now make it possible to carry through major operative procedures in the very young with equanimity Increasing knowledge of the physiology and pathology of the kidney and urinary tracts is guiding us to wiser treatment and more enlightened prognosis

Many of the disorders familiar to urologists in the adult are seen in their incipient stages in the child and their study in the young is therefore of great practical value and importance

With such a background the experiences gained over the past twenty five years or so are such that it is clearly a duty to record them for the guidance of those less fortunately placed, and in the interests of children everywhere

Such is the purpose of this book It is intended to be very much a practical guide to diagnosis and treatment, and its emphasis will therefore be found to be on the clinical picture presented by the child upon the investigations needed to establish diagnosis and upon the treatment to follow

There are still many uncertainties and unsolved problems in children s urology and it is hoped the experiences here set out may inspire others to help in furthering their solution

My own interest in this branch of children s surgery may be said to have been first stirred in 1912 by seeing Geo E. Waugh to whom we all owe so much successfully negotiating the passage of ureteric catheters in small children which at that time was no mean feat Thereafter interest quickened under the guidance and inspiration of my late colleague O L. Addison a pioneer in the urology of childhood to whose memory I gladly pay a very grateful tribute

It has been my happy lot to work at The Hospital for Sick Children Great Ormond Street, with its wealth of clinical material and traditional atmosphere of enthusiastic service and devotion I tender my sincerest thanks to all my colleagues there for the unceasing trust and loyalty which they have never failed to extend to me I have indeed been blessed with great opportunities

For the clinical work which provides the fundamental theme of this book I cannot evade personal responsibility but in its preparation I have been most fortunate in my two associates Each has provided a distinctive contribution based on personal researches particularly in relation to the relevant embryology

'suria

the infant, pain due to micturition frequently leads to retenti nmon causes of dysuria are meatal ulceration caused by "ammo ssibly associated with a meatal stricture

In the girl, congenital or inflammatory adhesion between the l imonia burns of the vulva are corresponding causes Apart from ises of cystitis and calculus, we have seen severe dysuria leadi nplicating the excessive use of sulphamerazine which forms a y : bladder (*see* page 53)

tention

already mentioned, complete retention of urine is most commo d spasm, and the impaction of sulphamerazine crystals in the ur ys has been encountered by us on 6 occasions These blockages itrumental dislodgement with a probe under anaesthesia Chror ine is fully dealt with later, but as a symptom it usually presen erflow, or as acute superimposed complete obstruction The ig periods between acts of voiding (for instance the child wl iter once a day) should lead to suspicion, and the measurem ine becomes a necessary part of investigation Bladder sensation m

continence

rinary leakage must be related to the normal development of u it broadly falls into 3 types

dribbling (day and night) or, in the tiny child, squirts of u minutes, this is often associated with chronic retention,

) the more or less involuntary voiding of larger quantities (2–3 c at intervals) with or without residual urine , this occurs in sever certain types of chronic retention,

i) the vertical incontinence, where there is a leak only when the but not when in bed, we have come to associate this partic feature with the presence of an ectopic ureter, though it does forms of "functional enuresis" in boys and girls

he quantity and concentration of the urine

i the urology of childhood, the interpretation of polyuria, oligu ust take into account the normal physiological variations wh iis period of life

It has been estimated that the normal infant during the first year i the urine about two-thirds of its fluid intake In the first few day scanty for various reasons, such as low intake, low blood pressur lood flow Fluid loss from vomiting or diarrhoea may reduce unces a day

The specific gravity of the urine and hence its colour is greatly reduced in the first year of life in contrast to the older child. This is a natural association of the infant s general immaturity shown by the inability of renal tubules to produce a markedly hypertonic urine (*see* page 28) The normal variations of specific gravity are so wide that special control is necessary if the test is to be of clinical significance In hot conditions the relatively large surface area of the child makes him particularly susceptible to excessive loss of fluid by the skin and children in such places as residential orthopaedic institutions exposed to the sun and air with regular controlled fluid intake are particularly prone to relative dehydration Despite these natural variations for which due allowance must be made observations of the urinary output and the specific gravity often afford valuable information

Haematuria

There is no need to emphasize the serious significance which must always attach to the passage of blood in the urine It must first be established that the source is in fact in the urinary tract Bleeding from vagina or bowel must obviously be excluded and this is not always easy in children

The passage of blood in the urine is rarely neglected Fear on the part of the mother usually means a very early investigation The classification of haematuria commonly employed in adult urology (painless and painful) is less applicable to the child especially to the infant Bleeding from a meatal ulcer is the commonest cause of haematuria in little boys The story is quite characteristic there being the usual pain and misery of the sore meatus scabbed over and then breaking open at each voiding. To this is added the alarm of a few drops of bright red blood at the beginning or end of micturition

Red urine of startling appearance may be due to extracellular pigment ingested in beetroot or dyed boiled sweets The clinician will not be deceived In genuine haematuria the blood component settles on standing and the presence of red blood cells is easily confirmed True haematuria may be accompanied by pain which may have its origin in the passage of a clot crystals or calculus, or in association with acute infections in which haematuria is seen not infrequently Most often pain is absent.

Serious haematuria usually painless, is a presenting feature of Wilms tumour more often than is commonly supposed Recurrent bouts of slighter bleeding are common in hydronephrosis and may also provide important evidence of renal tuberculosis Trauma may of course produce frank blood in the urine and it is well to remember that the recent administration of sulphonamides can be responsible

In the course of general diseases, such as the purpuras leukaemia and bacterial endocarditis haematuria is an expected incident, but it may be the initial mani festation The most important differential diagnosis however is from glomerulo-tubular nephritis in its initial stage Apart from the general features of the disease the presence of casts in the urine is usually confirmatory

Finally the rare forms of haemoglobinuria must not be forgotten Paroxysmal nocturnal haemoglobinuria, the Marchiafava–Micheli syndrome (Marks 1949) and other form of porphyrinuria congenital or acute which occur from time to time, must be watched for (Ashby and Bulmer 1950)

Cloudiness and pyuria

A parent sometimes observes that a child's urine is "thick" Most often the cloudiness is due to phosphatic deposit in the urine when it cools, but we have seen numerous children whose urine is cloudy with phosphates at the time it is passed Such a finding is of very great importance where there has been infection or stone, but in the otherwise normal tract this phosphaturia is unimportant It indicates lack of water intake and alkalinity

Turbidity which persists when the urine is warmed and acidified must lead to microscopic examination Frank pyuria is a common finding in children with dilatation anomalies of the urinary tract Sometimes the urine resembles pus and is too thick to be passed normally Ropy mucus adds to the difficulty The retention which occurs in bladders affected by spina bifida is a common precursor to such a condition

In the neonatal period, meconium passed from the urethra indicates the presence of recto-urethral fistula associated with the more severe type of imperforate anus

Pain

It is seldom easy to assess degrees of pain in childhood Considerable shrewdness and close observation are necessary Often there is only the history to guide us, and the version given by parents or other adults concerned may easily be exaggerated or the reverse Children themselves vary greatly in temperament and in their response to pain In infancy restlessness, whining and feeding difficulties may be indicative of a disordered urinary tract as they are of indigestion, and the two are of course often associated Severe pain is registered by the infant in screaming and kicking, which is perhaps best illustrated when micturition is attempted in the presence of a meatal ulcer

In older children the burning, scalding discomfort and dysuria, which characterize urinary infection, offer no great problem in recognition, but urinary tract pain of a more deep-seated character may be very misleading, since the child very commonly refers it to the abdomen

Tumour

Prominence of the abdomen due to bladder distension, the presence of hydronephrosis or renal neoplasm, often leads to the clinical presentation of a child in whom no other symptoms have been observed Routine examination of the abdomen in a sick infant may reveal the hypertrophic "cricket-ball" bladder of congenital bladder outlet obstruction, or, if distension has occurred, the bladder, as in the older child, may contain 20–30 ounces and be impalpable as a "tumour".

Effect of deterioration in renal function

Chronic ill-health, recurrent sepsis, anaemia and retardation of normal growth rate may each lead to routine investigations which in time reveal otherwise unsuspected urinary disease

Lack of growth can be related to urinary disease in several ways There may be a chronic uraemic state which leads to true renal rickets Renal function is to an unknown extent controlled by pituitary or other endocrine influence, and a disorder may simultaneously affect both renal function and body growth Chronic urinary infection and its toxaemia may, by a simple reduction of the child's vitality and appetite, be responsible for its lack of mental and physical growth

CHAPTER 2

CLINICAL EXAMINATION AND UROLOGICAL INVESTIGATIONS

THE chief symptoms with which we are concerned in urological cases have been described in the previous chapter but the importance of an accurate history regarding the onset can scarcely be over-emphasized and in the taking of the history one must form some estimate of the temperament and mentality of the child Defects of urinary control so common a feature of these ailments have obvious psychological repercussions on all concerned but especially upon the patient A sympathetic and confident approach is particularly important and brings its full reward in co-operation trust and gratitude

General examination

Due note is made of the child s physical development complexion height and weight. In many of these children signs of general ill health are surprisingly few On the other hand chronic urinary infection with the concomitant loss of sleep and mental worry is bound to lead to some deterioration of general vitality manifested in defective weight, lethargy and anaemia Serious renal deficiency will be evidenced in sallow complexion dwarfing and skeletal defects (renal rickets)

Clinical examination should be comprehensive particular note being made of the condition of the teeth nose and throat, chest, lymph glands and skeleton Upper respiratory infections (tonsillitis) have a considerable aetiological association with urinary infection A record of the blood pressure should be made as a routine as will be seen from Table 1 the child has a relatively low blood pressure and figures which may seem within normal limits for the adult may well indicate a mild degree of hypertension in the child In the examination of the abdomen the condition of the alimentary tract is of importance The intestine is the *B coli* factory and its significance in relation to urinary infections should not be forgotten Constipation has also much to do with disturbances of micturition and attention to this point will be well repaid

The lower pole of the right kidney is normally palpable in children and both kidneys are placed lower in the abdomen in the young infant than during later life Any undue enlargement, tenderness or tumour in the renal area must be noted The line of the ureter is carefully investigated for tenderness very rarely a grossly dilated and thickened ureter may be felt

The bladder in early life is vertically disposed and much more of an abdominal organ than in adults In children therefore the normal full bladder may be surprisingly obvious and palpable in the hypogastrium even when there is no question of retention The external genitalia are carefully investigated for any abnormality

A rectal examination often gives valuable information if only in confirming the existence of constipation The prostate is scarcely palpable as a definite structure

TABLE 1

Blood Pressure in Boys and Girls (after Nizzoli)
(Based on the findings in 2,278 children, as quoted by Fishberg, 1939)

Age	Boys		Girls	
	Systolic (mm)	Diastolic (mm)	Systolic (mm)	Diastolic (mm)
0–6 months	67	43	67	42
6–12 "	74	50	77	49
1–2 years	82	52	82	51
2–3 "	84	54	84	54
3–4 "	91	61	91	59
4–5 "	100	67	93	59
5–6 "	104	70	100	70
6–7 "	97	65	100	67
7–8 "	101	65	106	70
8–9 "	106	69	109	73
9–10 "	111	77	112	72
10–11 "	117	74	118	76

during the first years of life, but distension of the proximal urethra in some infravesical obstructions may occasionally be recognized Cysts of the utricle or even vesical diverticula may be palpable as mid-line swellings, and stones impacted in the lower ureter or in the bladder have also been detected by rectal examination

Urine

Needless to say, the urine should be examined immediately by the ordinary methods In most cases the assistance of the pathologist is not required to inform us of the fact of infection, though of course the bacteriology and drug sensitivity is an essential guide to treatment Bacteriological examination should normally only be carried out on a catheter specimen in the case of girls, but a clean specimen taken with full observance of aseptic technique is usually sufficient for boys The first few drops of urine should always be discarded even when passed through a catheter since they are liable to be contaminated from the urethra Twenty-four hour specimens may be required if the suspicion of tuberculosis arises In male infants urine specimens must be collected by the method of strapping a test-tube to the penis

In almost all in-patients it is wise to have a record of total fluid intake and urinary output

Renal function tests

In children one can usually form a fair estimate of renal function from general clinical observations before resorting to elaborate tests Blood urea estimations are valuable and are performed as a routine, though as an index of renal function they are apt to be falsified by dehydration from vomiting and diarrhoea, particularly in infants Readings over 40 milligrams per 100 millilitres may be taken as abnormally high The specific gravity test is simple to apply and often gives a

valuable indication of the degree of renal damage Urea clearance and concentration tests are seldom called for in surgical cases in which we tend to rely a great deal upon the intravenous pyelogram since this latter method gives an idea of the function of each kidney rather than an over all value

Excretion urography

Excretion urography is the most important single item in the investigation of the urinary tract and will normally precede any form of endoscopy since the information provided by the radiographs often suggests what must be sought by the endoscopist The intravenous pyelogram is a measure of renal function as well as an indication of the anatomy of the pelvis and calyces and must be viewed from both aspects The limitations upon its use are imposed by serious renal damage in which case the dye will not be concentrated sufficiently to cast a shadow and by the neonatal kidney which has not yet developed the concentrating power

In the young infant excretion pyelography may well present an insurmountable problem and under the age of 2 years it should only be attempted where the information cannot be gained by any other method The intravenous administration of the dye may well involve cutting down on a vein and there is much to be said for the intramuscular route using hyalase (1 milligram in each thigh) to expedite absorption Concentration is difficult to obtain in the infant and preliminary dehydration must be drastic It may also be wise to compress the ureters after the administration of the dye Since it is almost impossible to eliminate the gas from the infant s intestines clarification of the renal areas can be assisted by the administration of a Seidlitz powder which blows up the stomach with carbon dioxide

In older children the routine should be as for adult pyelography Fluid should be withheld for the preceding 12 hours in order to assist concentration and the bowel should be as empty as possible The elimination of intestinal gas is not usually achieved by the administration of aperients or enemas and it is wise to have the child out of bed and running about for 2–3 days before the radiological examination The solutions which may be used intravenously are shown in Table 2 It may be noted that the dosage given is higher than that commonly recommended by the manufacturers Only diodone solutions may be used intramuscularly

TABLE 2

B P	Boots	Glaxo	May and Baker
Iodoxyl (75)	Urumbrin	Pyelectan	Uropac
Diodone (35 or 50%)	Pyelumbrin	Pyelosil	Uriodone

Dose: 10 millilitres + 1 millilitre for every year of age.

Exposures are made at 5 10 15 and 20 minute intervals after the injection and after longer periods if secretion is delayed. These later skiagrams are often of the greatest value in outlining the dilated upper urinary tract Lateral views of both

kidneys and ureter may be of value, and it must be emphasized that in order to make the fullest use of this investigation there must be close co-operation between urologist and radiologist Excretion pyelography is not simply a routine procedure, but should be dictated by the urologist, under whose general supervision it should be carried out The timing of the later skiagrams should not simply be left to the radiographer but ordered by the house officer responsible after an examination of the wet plates of the earlier exposures

If and when the bladder is satisfactorily outlined by intravenous urography, a voiding cysto-urethrogram may complete the series and an estimation of the residual urine may be gained from a skiagram taken after micturition

For the proper interpretation of intravenous pyelograms it is necessary to have a clear idea of the range of normal variation, and this can only be gained by considerable experience of viewing these skiagrams The pelvis is in some cases narrow and spidery, with an elongated upper calyx in others it is large and entirely extrarenal The suspicion of cystic disease may arise in the first case, and of hydronephrosis *in the second, and only those accustomed to seeing many pyelo*grams can give a reasonable opinion on the borderline cases In children it is not uncommon to find that the whole length of the ureter is outlined in one skiagram and the lower lumbar spindle is often rather wider than the pelvic segment

Cysto-urethrography

The frequency with which vesico-ureteral reflux is a feature of urinary disorders in children makes the cystogram an unusually valuable investigation The use of sodium iodide as the opaque fluid is to be deprecated since it frequently has an irritant effect Diodone or iodoxyl solutions should be used at a concentration of approximately 10 per cent and it is usually convenient to dilute one of the more concentrated proprietary brands (for example Pyelectan 75 per cent 20 millilitres in 6–8 ounces water) The dye is injected through a catheter until the bladder is adequately distended and the child begins to complain The catheter is withdrawn and an exposure made The child then voids and further skiagrams, both antero-posterior and lateral, are taken during the act of micturition These voiding skiagrams are of the greatest importance since dilatation of the posterior urethra may be evident in them alone * In older children the density of the femora may obscure the urethral shadow in the true lateral, and oblique skiagrams are then required

Catheterization

In boys the introduction of a Jacques catheter (sizes 5–8E) seldom presents any serious difficulty provided that the manipulations are gentle and the lubrication adequate In cases of deformity of the posterior urethra, gum elastic catheters may be required In infant girls the urethral orifice is occasionally difficult to find and it should be remembered that the urethra is at first often directed backwards before turning vertically towards the bladder The catheterization in girls is considerably facilitated if, after the vulva has been cleaned with antiseptic, the labia are held apart with a dry swab which will not slip upon the moistened surfaces

* We are grateful to Douglas Stephens for impressing upon us the value of voiding cysto-urethrography

Residual urine

In infants with congenital malformations tending to produce urinary obstruction the slight trauma of catheterization may precipitate acute retention possibly with the introduction of infection In such cases therefore estimation of the residual urine should only be undertaken after careful consideration and should not be an out patient procedure In older children the dangers are not so great but care should be exercised We have found 1–2 ounces of residual urine to be not unusual in normal children

Cystoscopy

Although cystoscopy and retrograde pyelography are somewhat more exacting procedures, they are no less an essential part of urological investigation than in an adult. For the examination of the bladder we are accustomed to use a simple cystoscope with a fixed telescope which is available in a size approximately corresponding to F11 Single catheterizing cystoscopes of the Ringleb or Swift–Joly pattern can be made of approximately the same size but the difficulties of applying urological scales to instruments which are not circular in cross section and the optimism of manufacturers with regard to the delicacy of their own products, make it difficult to give an accurate calibration for all the instruments used The usual double catheterizing cystoscopes are about F14 The addition of an irrigating system slightly increases the size of the instrument and is seldom required for a child s bladder as bleeding is so seldom encountered In infant girls the examining telescope and single catheterizing instrument can be passed almost from the time of birth so that retrograde pyelograms are obtainable during the early weeks of life In older girls of course instrumentation presents very little difficulty The male urethra on the other hand exhibits a considerable variation in calibre and no definite statement can be made as to the age at which the various instruments may be passed On the average however the examining cystoscope can safely be introduced from the age of 6 months but one is seldom able to use the double catheterizing instrument until the age of 2 years or more At any age forceful instrumentation is dangerous and cannot be countenanced

A general anaesthetic is essential with very few exceptions and the fact that the child is unconscious demands special gentleness and care in every detail of the manipulations The child is placed in the dorsal position with the lower limbs flexed and abducted (*see* Fig 1) The parts are cleansed with cetavlon and the towels adjusted A metal catheter is then passed the bladder washed out and filled Metal bougies are passed up to the size of the cystoscope to be employed These bougies should be of a special child s pattern with a short and gentle curve as the fully curved adult instruments are very difficult to manage The filling of the bladder is best controlled with a hand on the abdomen and for the most part we prefer to use a syringe rather than the irrigator so that the exact quantity introduced may be appreciated The examining cystoscope is then passed for a preliminary inspection

The cystoscopic field is relatively small and the identification of features at first difficult but *experientia docet* The mucosa in general is relatively pale and the wall thin so that interlacing muscle bands may be clearly seen but this normal appearance must not be mistaken for the trabeculation of hypertrophy The

trigone in the child is less clearly defined than in the adult, the interureteric bar less evident, and the mucosa in fainter contrast The trigone is disposed somewhat more vertically and a *bas fond* is never observed The ureteric openings do not always show up very clearly and some patience may be required in finding them.

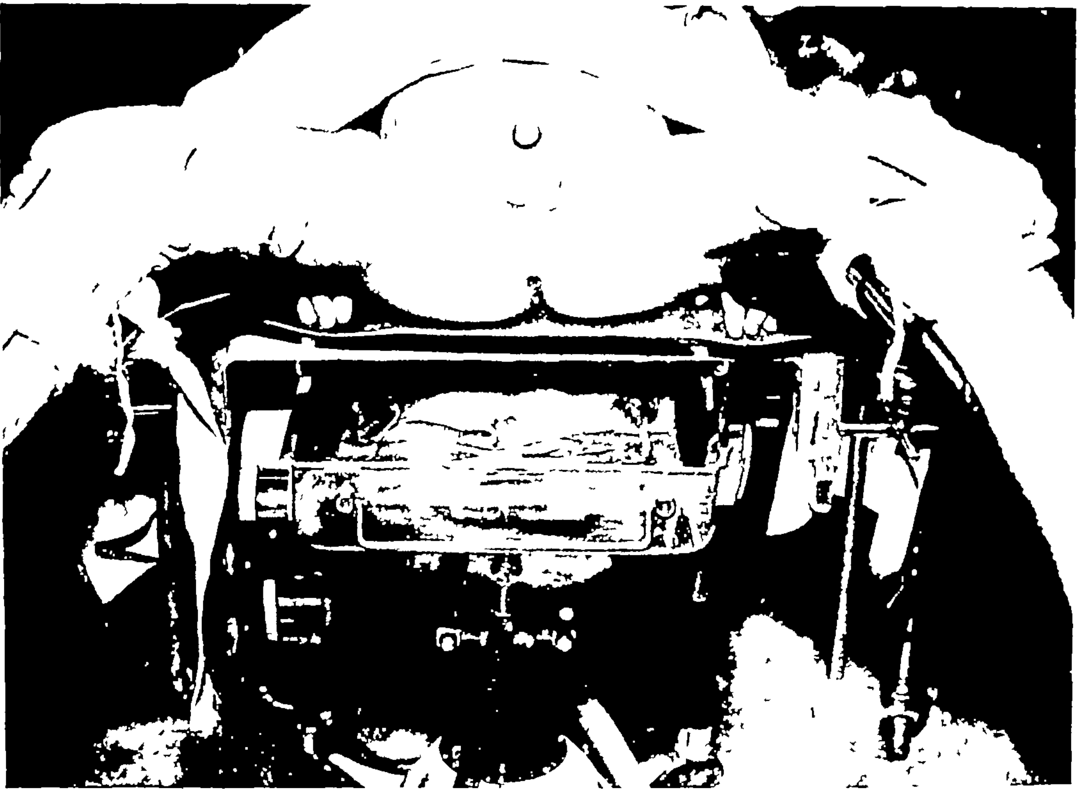


FIG 1 —Position of child for cystoscopy Operating table adapted by detaching movable section and fitting gutter knee supports The drainage tray runs out from beneath the table The head end can be tilted downwards if necessary

As will be seen in later chapters, we attach considerable importance to the appearance of the ureteric orifices, and in them often lies the greatest interest of child cystoscopy We have come to regard the injection of indigocarmine of little practical value in children's work, and in fact we hardly ever employ it nowadays

Ureteric catheterization

The passage of ureteric catheters is, as a rule, perfectly easy, but there are many circumstances in childhood which make for difficulty and on occasion one has to admit defeat The small-sized catheters required are apt to be soft and are checked by the slightest obstruction of a mucosal fold Retention of the stylet may facilitate the progress of the catheter but is apt to traumatize the wall Olivery tipped catheters are easier to pass but are in scarce supply and are liable to break Oedematous cystitis may make it difficult to detect the ureteric openings at all, and even when they are seen attempts at catheterization may well fail Many of the lower ureteric anomalies present obstacles which are difficult or impossible to negotiate

In passing the catheter the centimetre markings must be noted with care as the tip approaches the pelvis as it is not difficult to perforate the kidney We have experienced this mishap on 3 occasions though fortunately in none of them did the child suffer any inconvenience In very dilated ureters catheters curl up and return on their track and may even come out into the bladder again making pyelography impossible

With the catheters in position specimens are collected in rubber capped test-tubes attached to the child s thighs If the urine does not flow readily at once the catheter may be cleared by injecting and sucking back 1–2 millilitres of saline The differential specimens should be examined microscopically and the percentage of urea may also be estimated to give an idea of differential function but in our experience urine culture has not been of any value Organisms are very rarely grown even when the kidney is grossly infected and it seems that with these very small catheters, however carefully they may be washed through before use enough formaldehyde remains in them to sterilize the specimens

Retrograde pyelography

The child is taken to the x ray table while still anaesthetized or the skiagram is taken in the operating theatre if facilities are available A solution of 15 per cent diodone may be used, the amount required varying very greatly from case to case The normal renal pelvis holds approximately 1 millilitre of fluid for each year of life up to the age of 5 or 6 which gives a rough idea of the amounts which can safely be injected according to age If the preliminary intravenous pyelogram has given some idea of the capacity of the pelvis it is however better to judge from this than to follow any set rule No force should be employed during the injection since pyelovenous reflux will obscure the picture and rupture of a calyx may well be produced by unnecessary pressure The wet film should be inspected before the catheters are withdrawn and more dye injected if necessary Lateral skiagrams and views of the ureter filled as the catheters are withdrawn are often helpful

Urethroscopy

For the proper inspection of the posterior urethra a direct vision or foroblique telescope with irrigation is essential We employ 2 sizes of instrument, F11 and F13 the larger of which can carry a diathermy electrode or a ureteric catheter In the male the verumontanum and the orifice of the utriculus may be inspected and the condition of the vesical meatus and capacity of the proximal urethra noted Distension cysts of the utricle mucosal tags and polypi may be recognized and treated by diathermy The congenital valves which are so frequently responsible for urinary obstruction we have not found easy to identify urethroscopically and certainly they cannot be seen with sufficient clarity to allow their destruction by diathermy The bulging pocket on either side of the membrano-prostatic junction will often suggest the diagnosis but treatment, we believe is by operation

Anterior urethroscopy is seldom required although a small straight anterior urethroscope is available

Cystometry

This investigation consists of a simple measurement of the intravesical pressure during the slow filling of the bladder through a urethral catheter Its use reveals

the presence, absence or degree of sensibility of the bladder and indicates the automatic response of the musculature to distension In the past this simple procedure has been used mainly as a research method in cases of neurological disorder, and practically all that is known of the physiology of micturition is based upon cystometric investigations

Delayed maturity of bladder control may be evident from the cystometrogram, but in practice very little information is obtained that cannot be forecast from a careful scrutiny of the symptomatology or from cystoscopy Nevertheless we consider that the investigation will occasionally reveal helpful data in difficult cases and full details of the technique employed in children will be found in the Appendix

Summary

In conclusion it will be seen that a complete urological investigation involves many technical difficulties and is attended by not a few risks No child should be subjected to any part of the procedure for merely academic reasons or for the sake of routine Such information as is essential to accurate diagnosis must of course be sought, but once it is obtained let that suffice What is not absolutely essential should be avoided, however interesting it may seem to us

In practice it will be found that the affair often resolves itself into something of a " jigsaw puzzle ", the various bits and pieces having to be fitted together to obtain a composite picture

CHAPTER 3

PHYSIOLOGY OF MICTURITION AND RELATED DISORDERS

THE acquisition of the normal micturition control is dependent on many factors —the factors inherent in the individual in his training and his environment The dating of such control is thus liable to considerable confusion in the interpretation of what is normal and what is abnormal In older children who have been normally controlled and who then develop disordered micturition—retention incontinence frequency dysuria, urgency and so forth—the indication of abnormality is clear enough In the case of the infant or very young child it is wise to have a standard by which the persistence of excessive wetting or delayed control may be judged and its potential significance appreciated. Without such a standard delay in recognizing the abnormality may well be of serious consequence to the child

The physiological basis

Although during recent years experimental evidence has shown that the cerebral cortex may play a part in micturition beyond that of inhibition (Clark, 1945) the theory propounded by Denny Brown and Robertson (1933) has been basically unchallenged It is generally accepted that the primitive spinal reflex whereby the bladder when it becomes uncomfortably full is automatically emptied is the sole mechanism in the new born period With the passage of months this spinal reflex becomes modified to an increasing extent by inhibitory impulses from a higher level and eventually from the cerebral cortex. Whether or not there is an excitatory centre in the cerebral cortex is still undetermined but it seems probable that excitatory as well as inhibitory impulses may proceed from the brain Cortical disturbance might, therefore affect inhibition or excitation of micturition The field of inquiry now being traversed by electro-encephalography has opened up new vistas and evidence is accumulating which associates disturbances of micturition with particular areas of the brain and with associated disorders of function such as hypersomnia (Strõm-Olsen 1950)

Extensive cystometric study of the infant s and child s bladder control (Nash, 1950) has shown a consistent sequence in the pattern of response of the bladder at different ages This study also revealed a significant degree of correlation between the symptom stages of enuresis and the bladder behaviour during cystometry There appeared, therefore very strong experimental evidence to support what has for many years been held to be the basis of functional enuresis in children namely that it is mainly a condition of delayed physiological development

The development of toilet control

Functional maturation

McGraw (1943) has called attention to the very striking association between neural maturation and the training programme exemplified by the infant s

During the normal period of gesture indication of desire (10–20 months) a mother may have noticed a child's apparent discomfort or desire to void, but in cases of chronic retention the obstruction may by then have destroyed bladder sensation and any desire to void as indicated by grimace, gesture or sound may have gone.

After the age of 2 years accidents by day should be rare after 3 years they should be virtually unknown. Persistent and excessive wetting, particularly after the age of 2 years, should arouse disquietude since it may well indicate some serious abnormality of the urinary tract, but a child should not be considered as "enuretic" until after the age of 4 years

The cystometric picture

In the new-born infant sudden and powerful contraction of the bladder occurs quite automatically when perhaps an ounce of urine has accumulated within it As has already been mentioned, this reflex may be set off by some trigger action such as lifting the child or changing the napkin or even by distension of the stomach during feeding, and during these early months the cystometric pattern

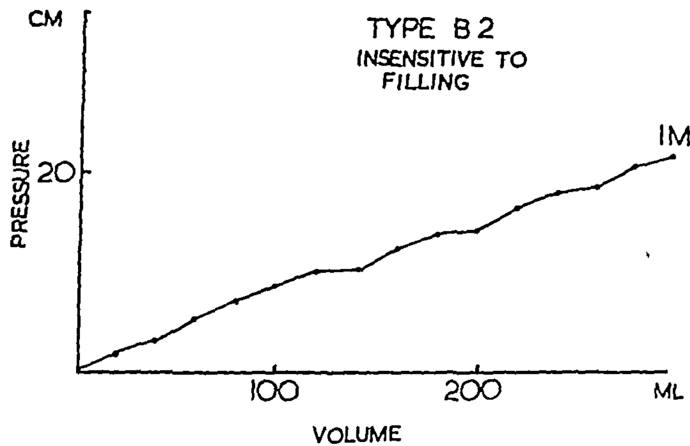


FIG 4—Cystometric graph from a boy aged 7 years, note absence of any intermediate points of "desire to void" This bladder, as a result of chronic infection and distension, was insensitive except to extreme stretching

is that of small capacity with one uninhibited contraction (Type A1 High tension, Automatic infantile) (Fig 3 (*a*)) During the latter-half of the first year, as the capacity of the bladder is increasing, contractions appear which are inhibited before voiding occurs The contractions are probably initiated by the same trigger actions as previously but inhibition from medullary or higher centres is coming into play (Type A2 High tension, Incompletely inhibited) (Fig 3 (*b*)) The child of 3 years presents a cystometric picture more nearly approaching that of the adult, where the base line pressure is lower and less steep Minor contractions reach consciousness but are inhibited and a considerable degree of volitional control over the terminal contraction has now been achieved (Type *B1 Low tension*, Incompletely inhibited) (Fig 3 (*c*)) In the adult pattern, which is often found in children of 5, there are no contractions whatever in spite of considerable discomfort

towards the end of filling (Type B2 Low tension Inhibited Adult) (Fig. 3 (*d*))

It is, therefore possible to get particular cystometric patterns associated with what may be called the normal bladder age

Cystometrograms taken on different occasions in the same child show a consistency of pattern and of abnormality and readings taken actually during sleep show that bladder contractions can occur and yet be inhibited before voiding takes place

It is convenient to give these cystometric types names which indicate the behaviour of the bladder Even the simple effort of deep breathing during investigation brings about inhibition and relaxation of the spontaneous contractions which occur in the partially inhibited types A2 and B1 (the cystometric types are shown in Fig. 3 (*a* *b* *c* and *d*))

Cystometry may reveal that the bladder is insensitive and passive to an abnormal degree as, for instance, in cases of long standing obstruction or infection (Fig 4) (For the technique of cystometry *see* Appendix II)

The clinical basis of enuresis

A developmental conception

Unfortunately many cases of urinary incontinence by day or by night appear first under the misnomer of enuresis and are found to be due either to retention with overflow to neurogenic lesions of the bladder or such gross abnormalities as ectopic ureter and it cannot be too strongly emphasized that organic disease of the urinary tract must be excluded before a child is said to have functional enuresis *

There are three big groups of cases First those in which there has been a delay in or a disorder of control of micturition from birth If the expected variation in control during the first 3 years of life is borne in mind the clinician will not be misled by the mother who asserts that her child became dry at 6 months. Children who have shown in the first 2 years that they have the mechanism for controlling micturition by being dry for 3 or 4 months often get swept back in the ebb-tide when they begin to talk and take a wider interest in their surroundings

Secondly there are those children who have achieved perfectly satisfactory control both by day and night within a reasonable period say by the age of 4 or even 5 years, and then after a period of perhaps years have relapsed to the infant state of being wet at night and perhaps during the day as well These constitute an onset group

Thirdly there are those children whose night control is normal but who are wet during the day This last group is a small one and one in which there is often in our experience, either a gross organic disease or very gross psychological disturbance This group is not in series with the previous two and is a distinctive pattern, not related to physiological maturity in the same way

Inquiry into the history of a particular case should ascertain in addition to the usual facts (such as birth weight, obstetric difficulty neonatal asphyxia, jaundice

* The term enuresis strictly refers to making water in the bed from the Greek ενουρεω The French *pis en lit* (dandelion) is the equivalent, but the term has become hallowed by use and is applied only to those cases of urinary incontinence in which no organic structural cause can be found

or initial feeding difficulty), whether the child has actually had a single dry night at any time in its life. If there has never been a dry night, then an attempt must be made to determine what is the greatest number of hours the child has been found to be dry It is our experience that such a child has always had day symptoms such as marked frequency, urgency and even wetting In the distress of the condition parents have very often ignored the day symptoms If there has been a dry night at some time or a succession of dry nights, then the parent must be asked what is the greatest number of nights in succession that the child has been dry, "has it been weeks or months," and "what is now the average number of wet or dry nights a week?" An inquiry should be made as to the depth of sleep or whether the child is restless in bed If the child is "lifted" at the parents' bed-time a note should be made as to whether the child is in fact dry then When a case falls into the second big group, known as the "onset" type, any factors which may have led to the return of the infant state must be sought for A severe attack of whooping-cough, a physical injury, change of home or change of school are all factors which we know are important and which will be discussed later

Upon the basis of clinical history we have found that all but a very few cases of enuresis fall into one of several symptom groups which are surprisingly consistent and which correspond with the phases found in the development of normal micturition control These stages are described in simple terms and form a definite basis of assessment of the condition and progress under treatment (Table 3)

TABLE 3

Stage	Symptom group	"Bladder age"	Common cystometric type
I	Lifelong night wetting Never had a dry one Day symptoms	0–1 years	A1
II	Lifelong night wetting Occasional dry one Day symptoms	1–2 years	A2
III	Lifelong night wetting Remissions up to a month Day normal	2–3 years	B1
IV	Remissions over 1 month Day normal	3 years	
V	Very occasional wet night		B2
VI	Normal		
	Onset type		Mixed

Stage I This represents the persistence of the first-year physiology, and within this group the important further point is the severity of day symptoms

Stage II This state is similar to Stage I but the occurrence of one or more nights has shown that the mechanism of bladder control is intact even if it is turbed in its function We have been accustomed to take as an empirical divi line between Stage II and Stage III an average of 3 dry nights a week and presence or absence of day symptoms It is uncommon to find that a child has day symptoms has more than 3 dry nights a week on an average and w the child reaches the stage of having a week dry at a time he very rarely has symptoms The significance of this distinction between the Stages II and III be emphasized again under the discussion on treatment.

Stage III Children who have reached the stage of remissions of a wee more have usually lost their day symptoms and this group forms the most diffi the largest and the most disappointing of all the cases of enuresis

Stage IV This is represented by the child who has perhaps 2–3 wet nigh month and whose enuresis is a worry to the parents but not a severe disabi although it may interfere with schooling in the case of a boarder

Stage V One or two wet nights a term usually represent the residue of a l standing weakness and for such wet occasions it is usually possible to put o finger on an incident such as a school play or an outing which by leadin over fatigue and excitement has produced the nocturnal disaster Cases in stage do not really constitute a clinical problem

Cases of the onset type have shown a relapse to a primitive state which be assessed from the symptoms as being in one of the first 4 stages and theref for descriptive purposes, such a case can be labelled Stage II (onset) instance Such an annotation gives from the notes a very clear picture of condition when the child is first seen

There will be occasional cases which cannot be fitted into one of these gr but we have found the incidence of such cases to be less than 0 5 per cent, the exclusion of those who are wet by day only Table 3 shows the correla between the symptom grouping and the average cystometric finding. It mus emphasized however that by no means all of the cases in any particular s will show the appropriate cystometric pattern but the majority of cases will d

Aetiological factors in enuresis

There are many factors which are known to play a part in this delay in phy logical maturity which shows itself as functional incontinence In a series of consecutive cases referred to 3 clinics purely on account of the symptoms descr as enuresis one of us (Nash 1950) found that in 86 per cent there was known cause whilst in the remaining 14 per cent, 10 had gross unsuspected org disease of the urinary tract and 4 had other organic disease which could be sidered as the direct cause (such as cerebral palsy petit mal) In the cause known group (86 per cent) 20 per cent had past or present urinary tract les 8 per cent had moderate or severe threadworm infestation and only 58 per were without any physical abnormality Of the total group referred for treatm of enuresis, therefore 42 per cent required medical or surgical treatment for o lesions, either causing or associated with the enuresis

Constipation is quite clearly an aggravating factor in the maintenanc functional urinary incontinence especially in those cases described as havi

"terminal reservoir" in the colon In such cases, rehabilitation of the colon often leads to a complete disappearance of urinary symptoms

Posterior urethritis and trigonitis are said to be responsible for a certain number of cases of enuresis, and Winsbury-White (1944) found that 70 per cent of cases had urinary pathology We have not examined all the cases endoscopically and do not consider this to be an essential early investigation, though posterior urethral changes such as polyp and montanal oedema may be found they are rarely an important causative factor Meatal stenosis and meatitis, adherent prepuce, vulvitis, scabies, hernia, hydrocele and undescended testis are all conditions which may aggravate enuresis or perhaps, more fundamentally, delay the development of the normal degree of cerebral control over an inherently excitable organ It may be found that as a result of encephalographic studies specific cortical lesions or abnormalities of rhythm are responsible for a certain number of cases of enuresis, and the association of the very troublesome delay of micturition control in cases of organic nervous disease in children is well known In the "onset" type, sunstroke, whooping-cough, bombing incidents (in which the child has sustained blast injury), asphyxia and encephalitis have all occurred as precipitating factors

Something has been said already of the association of deep sleep with night incontinence, and whether the incontinence is the result of the deep sleep or whether both are the result of some underlying hypothalamic or cortical disturbance is undetermined (Strom-Olsen, 1950) Many children sleep very heavily but in spite

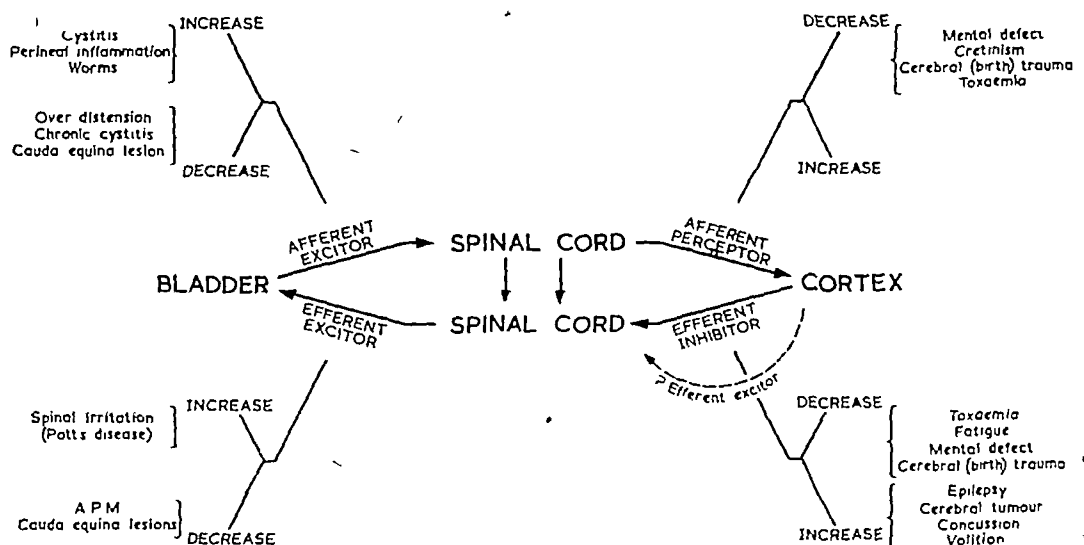


FIG 5 —Factors influencing the control of micturition

of this have complete bladder control It is doubtful if the fact of deep sleep is actually contributory but treatment directed to lighten the sleep level is very often effective

Undoubtedly heredity, housing, social environment and discipline all play a very great part in the training of a child, as much in his bladder control as in his table manners Inquiries into these aspects of the condition are all within the ambit of the physician and surgeon and we have seen little, if any, benefit accrue from

the reference of children suffering from enuresis to Child Guidance Clinics unless the urinary symptom is the least disturbing feature of a gross behaviour problem

The aetiology of this condition is still when everything has been said nebulous to a degree which is exasperating Fortunately treatment is more successful than attempts to elucidate the cause.

Fig 5 indicates some of the postulates in the aetiology and therefore in the basis of management of disorders of micturition control

This disturbance of function is sometimes a distressing residue after the completion of extensive surgery in the urinary tract even in the absence of any persistent organic lesion

The management of enuresis

General principles

The management of urinary incontinence in the child is not merely a urological problem, neither can it be circumscribed by any other specialist department of medicine or surgery. Having its basis more often than not in disordered physiological development and being accompanied either as a causative or resultant factor by urological or other disorder this inability of a child to hold its water according to the normal social pattern expected of children of similar age must demand more than the fleeting attention of all those who have to do with children in a medical sense Treatment must of course be pursued along aetiological lines It is not intended here to present a complete survey of all the many methods which have been claimed to cure certain cases of enuresis, but to point out the line along which we approach a particular problem so that the maximum result may be obtained in the minimum time Cases which come to a clinic which is primarily urological tend to be those who have failed to respond to simple measures and include those who have become the despair of the Child Guidance Clinics

Whatever line of treatment is pursued first there are always social and environ mental factors to be discussed and perhaps adjusted The outlines of treatment discussed here apply equally to those cases of life long duration and those where the enuresis has come on after a period of years or months of control

Social environment

The parental attitude

It is important that the parents should be encouraged to adopt an essentially practical attitude to the condition and not in any way to despise the child or them selves for this apparent failure to achieve some degree of social maturity They should maintain a firm attitude of discipline but not of punishment They should be sympathetic but not tolerant of the unpleasant state of affairs The most difficult relationship is usually that of the boarder away at a preparatory school where the parents reject all sense of responsibility for the child during the term time Perhaps the most vital thing to impress upon parents is that regularity in treatment and routine is a matter for which they themselves must be entirely responsible Children have very little sense of time and cannot be expected to remember their treatment or their routine without some prompting. Week-enders are very bad parents when it comes to dealing with enuretics Similarly parents who visit the

speedway or the greyhound track too many nights a week must be told frankly of their own part in the development of the condition A clue to this background very often comes from the fact that the child is wearing a cinema club or speedway badge and the clinician should be on the look-out for such things, which will never be presented to him as direct evidence

The domestic facilities and general hygiene

Housing difficulties in the last decade have made it extremely difficult for many parents to train their children in a satisfactory toilet routine Many of the cases of enuresis which developed during war-time evacuation were due to the absence of toilet facilities within the house Whatever may be the psychological cause, the association of an incontinent child with another child of the family by sharing a bed or a room helps neither the sufferer nor the observing child and it is not superfluous to quote practical examples of the "contagious" nature of the condition A boy of 9 years, with urinary incontinence due to spina bifida, during the school holidays had to share, for the first time, a room with his younger brother of 5 who had by this time attained perfect control the younger boy one morning noticed the elder boy's bed to be soaking wet, and having commented thereon to the elder boy proceeded to wet the bed every night himself for some weeks until a very firm line was taken by his father Another boy, despairing of his own enuresis, ensured that his elder brother should suffer the same degradation by pouring the contents of the chamber into his brother's bed each morning A bedside torch, a light on the landing, a lowering of the catch on the closet door so that it comes within reach of the small child's hands, are all practical points which help in certain cases

The nature of the bed and its coverings also play a part by altering the level of sleep It has often been noted that a child is dry for a period away from home, for instance, when sleeping on a palliasse at a scouts' camp, and the substitution of an ordinary hard hair mattress or a camp bed for the luxuries of a sprung interior mattress may achieve an immediate cure Less blankets on the bed may also be a help This over-protection of the child at night is very often the result of misdirected parental concern Warm but light coverings and an open window should be prescribed specifically Conversely, too little protection at night may be an aggravating factor

Reference has already been made to the significance of constipation, and in the child who is suffering from severe stasis in the lower bowel it may be well to advocate colon wash-outs at the beginning of treatment to make sure that there is really an effective evacuation of the bowel It is no use attempting other methods of treatment of functional incontinence when the rectum is continually loaded with hard faeces

The child's responsibility

All children coming for treatment should be told firmly but kindly by the doctor that they really must help themselves in the treatment, and that he will expect them to do certain things, particularly in regard to keeping a calendar It is a reasonable thing also to request the parents to see that the child removes from the bed in the morning any dirty linen and puts the sheets into soak as part of the routine on occasions when they have been made wet Once the child can learn to

tell the time he can be made responsible for regular visits to the toilet during the day at whatever time-intervals are prescribed and a very firm attitude should be adopted towards the persistent day wetter

The day symptoms

With that general background of guidance attention can now be turned to the symptoms which persist during the day and the parents can be told quite definitely that they are very unlikely to succeed in persuading the child to be dry at night consistently as long as there are any day symptoms The main focus must be on this day frequency and if there is dampness or wetting it is very unlikely that there will even be one dry night First we would emphasize the futility of trying to persuade a child to wait when the bladder has already called out full and the only case in which it is permissible to tell the child not to go to the toilet and firmly to prohibit its doing so is the particular type in which the child between going to bed and going to sleep perhaps over the space of an hour and a half may wish to visit the toilet 6–7 times This is a rare but very definite clinical entity which can be overcome by firmness and the administration of phenobarbitone at tea time Otherwise the underlying principle of treatment which has been found to be almost infallible is that the child must be sent to empty the bladder so often that he never experiences the desire to go This avoidance of the afferent impulse certainly seems to work and the dramatic abolition of life long day symptoms occurs in the space of 48 hours in many cases Its success depends entirely on the exactitude with which the method is explained to the parents and on the faithfulness with which they carry it out. When one starts to explain this system to the mother literally without exception one is greeted with the retort, I cannot do that because he goes to school One s reply must be that the procedure must start on a Saturday morning and the child must be kept at home. He must be made to pass his water at 8 o clock 8 15 8.30 8 45 and so on or perhaps starting at half hour intervals if it is found that he can manage this This voiding at what may appear to be absurdly short intervals of time must be done by the clock At lunch time the interval is extended to three-quarters of an hour if he has achieved half an hour dry by then and is continued at three-quarters of an hour for the rest of the day No particular care is taken at night and the child should not be lifted more than the once at 10.30 or 11 On Sunday morning the routine is recommenced still by the clock at three-quarters of an hour intervals, extended at lunch time to an hour If it is found that the child experiences a desire to pass its water in the intervals then the timing must be set back by a quarter of an hour and the procedure repeated for a half day It is not an over-statement to say that in very few cases does this fail to achieve an absolute control, so that by Monday morning the child can set off to school knowing that he can be dry for an hour and with strict instructions that he must go and pass water each hour without fail With the child who cannot remember or who is too young to know either he must be kept at home or a note must accompany him to school with an explanation for the school teacher It is regrettable that the co-operation of school authorities is still deplorably poor and the majority of mothers are told by the school authorities that they cannot fuss with children They sav that the child is allowed to go out if he wishes, but the whole point of this training is that the child

does not know until it is too late and he must be sent at set intervals and not merely allowed to go when he wishes Over-packed classes, harassed and sometimes ill-trained school teachers all contribute largely to the persistence of the condition of enuresis in our big cities

When the hourly system has continued for a week, an attempt may be made at the following week-end to increase the interval by quarter-hour periods, and even when the 2-hour interval is reached the parents must be warned that this frequency must be maintained by the clock and under no circumstances should the child merely be allowed to wait until it wishes to go to empty the bladder Many parents are so encouraged by the result of this very simple training that the whole attitude in the household changes in a matter of days This system of "time-training" is used by us as a routine in children admitted to the hospital for investigation of the condition, and there are very few of such children who do not acquire sustained control during the short period of their admission The same procedure of progressive training must be adopted if the child should relapse at a later date

The common thing is to find that after 4–5 weeks of this consistently good day response the child starts on his own to have dry nights when he has not already done so before

On general principles it is very much better not to inform the school of the condition, for the child's sake, unless no response has been obtained by the mother's efforts alone in training the child The system of "time-training" is of the utmost value in institutions such as orphanages, where the laundry saving very quickly repays the effort required to ensure periodic visits to the toilet

If a child fails to respond appreciably to this system, then we would emphasize the necessity for early, complete urological investigation, and such a child will most likely be found to have a bladder which, on cystometry, shows a persistent high-tension response with very small capacity, such cases over the age of 6 years are extremely difficult to treat.

The night routine

There seems no particular virtue in restricting the fluid intake before going to bed but such diuretics as tea and cocoa should be avoided It is imperative that the child should be woken and made to empty the bladder about 3 hours after going to bed. If the child is wet, then the interval must be reduced so that at least the first part of the night is dry Not until the day symptoms have gone and this early part of the night has been found to be consistently dry will full night control be established Parents have often asked if they should lift the child again during the night, but it is much wiser to leave the child completely undisturbed till the normal rising time Only those children who have failed to respond to the usual methods of treatment should be woken during the night by the parents or by an alarm clock

Sleep levels

Reference has already been made to the level of sleep as an aetiological factor, and to the association of hypersomnia from fatigue or toxaemia with nocturnal urinary incontinence The hardness of the bed, the scantiness of the bedclothes and avoidance of over fatigue during the day are all factors which lighten the level of sleep Whether a child wakes up when the bladder is full or continues to sleep so that the bladder empties itself automatically depends upon the intensity

of the afferent stimulus, the condition of the cortical receptor and the intensity of the subsequent inhibition impulse It is possible by various means to increase the responsiveness of the cortex or whatever nerve centre may be involved, assuming that there is a normal afferent impulse from the bladder (*see* Fig. 5) Considerable success has been claimed though we have little personal experience of it, for hypnosis as a method of increasing this conditioned response Intravesical tension is increased during hypnosis by manual pressure on the suprapubic region when the child is given the necessary instructions Hypnotic treatment is of course, based on an entirely different principle from the majority of psychological treatments which are more analytical in their nature and assume that the enuresis has not a developmental or physical basis but is due to some complicated psychological disorder Hypnotic treatment is much more rational in our opinion The use of the electric pad is undoubtedly successful in other cases where lack of response to the bladder filling due to deep sleep is important. Davidson and Douglass (1950) have described the apparatus and its use. The principle is that as soon as the micturition starts the electrode on which the child is sleeping becomes wet and the 2 layers of copper gauze which have been separated by lint are short-circuited This works a relay which in turn sets off an alarm bell which continues to ring until the child gets out of bed and turns it off Thus the bell rings a very few seconds after the bladder has given the signal full He then empties his bladder completely and consciously

The most simple and most effective way of ensuring that the maximum use is made of the minimum response of the bladder is to use sleep-lightening drugs, of which dextro-amphetamine seems to be the most effective For many years ephedrine has been used in the treatment of enuresis and it may well act by damping down the automatic contractions of the bladder which occur when it fills but it is more likely that its effect is on the higher centres by raising the level of sleep It is of course well recognized that the over-dose symptom of ephedrine is restlessness at night We have used methyl-ephedrine which has a longer excretion time very extensively with considerable success, but on the whole we favour the use of dextro-amphetamine When the child is over 5 years, this drug is given in the dose of 2 5 milligrams to start with at bed time, and if the child is dry when lifted at 11 p m or thereabouts the dose may be repeated at this time and increased by 2 5 milligrams at a time until the maximum tolerated dose is reached. Symptoms of over-dose are restlessness, nightmares and a feeling of intense fatigue the following day usually shown in a child by bad temper and listlessness A particularly anxious and worried child may safely be given 2 5 milligrams in the morning as well for an entirely different—and some might say perhaps unjustified—reason but, nevertheless, in practice the increased confidence which the child obtains more than outweighs the risk Some children of perhaps 8–9 years will tolerate 10 milligrams of Dexedrine at bed time and still sleep right through the night without any disturbance whatever Such a dose would keep the average adult awake the whole night This excessive tolerance of dextro-amphetamine is an indication that the treatment is proceeding along the right lines

Other therapeutic measures

The use of belladonna requires little comment and we would say that the only place for its exhibition is in children under the age of 5 years where it may be given in

pill form as the dry extract of belladonna $\frac{1}{2}$ grain at night Methyl-ephedrine has already been mentioned, and at the age of 5 the dose should commence at $\frac{2}{3}$ grain at night and in the morning The only other substance that we have found of value is stilboestrol The sodden vaginal epithelium of a little girl with vaginitis responds very well to 10 days' treatment with 0 5 milligram stilboestrol daily, and this may contribute to reduction of the enuresis The same drug in the same dose given to the older boy may be effective in reducing priapism which is sometimes a very potent factor in the production of night wetting There is no risk in using this substance for perhaps 2 short courses in the dosage stated, or rising to 1 milligram at night in the older boy

Investigations—their timing and effect

A big question in treating enuresis is to decide at what point full urological investigation should be initiated and, when this has been done, in attempting to decide, if there has been an improvement, whether it is the investigation itself or the subsequent treatment which has brought the improvement about If after 3 months' initial treatment along the lines already suggested the child is not materially improved, we would suggest that a full urological investigation is essential Clearly, before the diagnosis of enuresis has been made, stools will have been examined for parasites and any infestation will have been cleared Any gross urological or genital abnormality will have been dealt with, and it is assumed that only those cases with no obvious organic lesions or in whom these lesions have been eliminated are now under consideration Intravenous pyelography should be the first major procedure, followed by catheterization The measurement of residual urine, if any, bacteriological examination of the specimen removed, and cystometry may then be performed Cystoscopy and urethroscopy may be performed on the same occasion or left until the effects of the passage of the catheter have been noted It is remarkable how often the child who has never been dry previously will have several dry nights after the passage of a catheter, even in the absence of any infective urethritis Irritation of the urinary passages raises the intensity of the afferent impulse and the muscular control of the bladder neck We believe that it is quite likely that the success which has been obtained through the passage of sounds in the past is due to this irritant factor We have also noted during cystometric investigations on a large scale the very great number of children in whom the enuresis is improved considerably for a period after the investigation has been completed Nevertheless, urethroscopy and cystoscopy should be undertaken to make a thorough search for the rare and hidden causative factors such as polyps of the posterior urethra, bladder-neck abnormalities, ectopic ureter and ureterocele The discovery of a structural abnormality does not necessarily mean that the cause of the enuresis has been found The removal or correction of the error will not always remedy the incontinence

Operative treatment

When all the organic causes have been eliminated and accepted methods tried, there still remains the "hard core" of cases who have not responded to treatment In our experience this amounts to far less than the 8 per cent quoted by other authorities (British Medical Association and Magistrates' Association, 1948)

We believe that the electro-encephalogram will unravel some of these problems The use of penile clamps, rubber urinals and other appliances has been resorted to in extreme cases We have had experience of a limited number of cases in older boys and young adults in which we have used an adaptation of the method described by Lewis for the operative treatment of urinary incontinence following prostatectomy (Lewis 1949) This operative procedure is described on page 127 It consists of placing within the bulbo-spongiosus muscle mattress sutures of silk which enfold the bulbous urethra so limiting the diameter of its lumen A reduction of the diameter of the bulbous urethra may enable the rather feeble bulbo-spongiosus muscle to occlude what remains of the lumen and act as an additional sphincter Whatever may be the explanation we have had several cases in which the cure of the enuresis has been dramatic We have also had others who have failed but in the successful ones previous cystoscopy instrumentation and anaesthesia have had no effect whatever in reducing the frequency of incontinence whereas the operation was dramatic This is therefore a method with uncertain success but worthy of a trial in otherwise unresponsive cases in older boys

CHAPTER 4

DAMAGE TO THE RENAL PARENCHYMA

Physiological considerations

THE diseases which affect primarily the renal parenchyma are not for the most part amenable to surgical treatment, but the urologist's preoccupation with the urinary passages must not be allowed to conceal the fact that the disorders with which he deals threaten the life of his patient chiefly through their secondary effects upon the nephrons In all cases of obstruction and infection of the urinary tract the renal function must be taken into consideration, and treatment of the biochemical disturbances is sometimes more urgent than any other measure

Normal physiology

The normal physiology of the kidney need not be described in detail the basic processes of glomerular filtration, tubular re-absorption and tubular secretion are accepted and familiar, while many of the detailed mechanisms are still a matter of controversy among physiologists The kidney regulates the composition of the urine in such a way as to maintain the constancy of the " internal environment ", and exhibits in health a remarkable elasticity in its capacity to meet the emergencies which arise from disturbances of the *p*H of the blood and of the water and salt balance A loss of this elasticity, with an inability to form a concentrated urine, is one of the first signs of renal failure Disturbances of the acid-base equilibrium are common in childhood and the part played by the kidney in their correction is paramount The end-products of metabolism are chiefly acid so that the urine needs to have a considerably lower *p*H than the blood This is achieved in part by an adjustment of the proportions of acidic and of basic phosphate with re-absorption of the fixed base (Na, K, Ca or Mg) as bicarbonate ($Na_2HPO_4+H_2CO_3=NaH_2PO_4+NaHCO_3$) The tubular cells also manufacture ammonia from glutamin and the amino acids which can replace the fixed bases

The vascular arrangements within the kidney are of considerable interest The work of Trueta *et alii* (1947) has made it clear that in the experimental animal there is a mechanism by which the blood is diverted from the glomeruli in the outer layers of the cortex and passes through the wide channels of the juxta-medullary zone The stimuli which operate this vascular redistribution are many and various, but there appears to be no doubt that, in severe shock, anuria may result from this diversion of blood There is every reason to believe that a similar mechanism exists in man, and that it plays an important part in the anuria associated with shock and the crush syndrome Anoxia may be an effective stimulus and in the human child Govan (1949) brings evidence in support of a diversion of cortical blood in neonatal asphyxia Nash (1951) has suggested that even in non-fatal cases birth trauma, resulting in cortical ischaemia, may be responsible for renal damage which will only become evident at a later date

In recent years McCance (1950) and others have drawn our attention to the considerable differences between normal renal physiology in the infant and in the adult, differences which might indeed be expected if the kidney is regarded primarily as a regulator of the internal environment The extracellular fluid of an infant represents some 50 per cent of its body weight as against 20 per cent in the adult the osmotic pressure of the plasma is the same but it contains less protein and more chlorides and phosphates The distinctive infant renal physiology must be regarded as an adaptation to these differences in the composition of the body fluids. The structure of the kidneys is immature in the infant Up till birth the glomerular loops are matted together and surrounded by a sac of high columnar epithelium (Gruenwald and Popper 1940) This epithelial sac disintegrates after birth and allows the loops to expand but the high columnar epithelium persists for some time The thick glomerular membrane resists therefore the normal process of filtration and the rate is very low compared with the adult figures The actual number of nephrons present at birth is supplemented during the first few weeks of life Tubular function is also immature during the first months of life both from the point of view of re-absorption and of secretion The diodone clearance in the new born is only some 10 per cent of the adult figure so that it is easy to understand why such poor pyelograms are obtained Urea clearances are low but under normal circumstances the total nitrogen output is low because of the rapid building up of proteins

The clearance of chloride and phosphate is lower than in the adult, a fact related to the greater volume of the extra-cellular fluids A low phosphate excretion however handicaps the infant in the maintenance of a constant *p*H and the infant s kidney has not the capacity for varying the concentration of the urine possessed by the adult organ Consequently if a large amount of urea has to be cleared from the blood a large amount of water and electrolytes must go with it, and biochemical disturbances are not rapidly corrected

Acute renal failure

When there is an abrupt cessation or an extreme depression in renal function the products of metabolism accumulate within the body the blood urea rises steadily and uraemic symptoms appear after a few days Unless there is a recovery of function death will inevitably occur usually 1–3 weeks after the onset of the disease Hypertension is not common in acute failure and only moderate in degree Oedema may result from excessive fluid intake and pulmonary oedema is a frequent cause of death Accumulation of potassium as a result of continued protein metabolism may be responsible for the sudden cardiac arrest. Depression of the serum chloride level is seen with severe vomiting, and acidosis is not uncommon

The great majority of lesions causing acute renal failure are in fact recoverable and although the mortality rate is at present very high, careful treatment should make it possible for many patients to weather the biochemical storm

Types

The lower nephron nephroses—A somewhat heterogeneous group of lesions is included under this heading, but the majority have in common an initial episode

of shock and a predominence of pathological changes in the distal convoluted tubules The "reflex" anuria, which follows operative shock or extensive burns, the "crush" *syndrome, the transfusion kidney and the post-abortion anuria may* all be taken as examples of the lower nephron nephrosis, though none of them is common in childhood It seems probable that the initial episode of shock is responsible for diversion of the blood into the juxta-medullary zone, leaving the cortex ischaemic (vascular shunt) The tubular cells are damaged by this ischaemia so that when the blood is returned to them they are unable to carry on their normal function, particularly if haemoglobin or myohaemoglobin needs to be excreted Thus after recovery from shock the patient is often well but has a diminishing urinary output which may cease altogether after a day or two The little urine passed is of low specific gravity and is frequently blood-stained Recovery of the kidney commonly begins about the twelfth day with the excretion of a gradually increasing quantity of very dilute urine, but the danger to the patient is not yet past, for the diuresis involves extreme disturbances of the water and salt balance, which may be sufficient to overwhelm an already dangerously ill patient The kidney in the lower nephron nephroses is usually swollen and oedematous and bulges out whenever the capsule is incised, a feature which has encouraged the operation of decapsulation Microscopically the glomeruli are often normal, the tubular epithelium is flattened and casts, often pigmented ones, are found in the distal convoluted tubules There are also characteristic tubulo-venous thromboses (herniation of casts into thin-walled veins)

The toxic nephroses —Mercury, phenol, carbon tetrachloride and the sulphonamides may be responsible for acute toxic necrosis of the tubular epithelium Particularly in the proximal convoluted tubules, the cells are swollen and converted into granular masses which may obstruct the lumen The glomeruli are comparatively unaffected, but small foci of necrosis and desquamation of the capsular epithelium may be seen As in the lower nephron nephroses a period of oliguria precedes complete anuria, the urine being cloudy and loaded with albumin and casts It is believed that the anuria is the result of complete re-absorption of the glomerular filtrate through the necrotic tubular epithelium The clinical picture is apt to be complicated by other manifestations of the toxin, for example mercurial colitis, but the progress of the uraemia is the same as in the other causes of acute renal failure Recovery with regeneration of the epithelium is possible and again diuresis may be responsible for a fatal biochemical disturbance

Sulphonamide anuria.—The cessation of renal function, which results from sulphonamide therapy, is a complicated process in which at least 3 possible factors may be operative obstruction, toxic necrosis of the tubular cells, and allergic changes within the kidney The acetyl derivatives of many of the sulphonamides, particularly sulphathiazole and sulphadiazine, may crystallize out within the urinary passages and obstruct them Such obstruction may occur in the renal pelvis or at the lower end of the ureter where it may be reached and relieved, but it may also occur within the renal tubules In many fatal cases, however, there is unquestionable cellular damage, and a picture somewhat similar to that seen in the lower nephron nephroses, with which sulphonamide anuria is sometimes grouped It is more probable, however, that a direct cytotoxic effect of the drug

is the responsible factor Occasionally renal damage accompanies general manifestations of allergy to the drug, and the kidney may then present a very striking picture There is some fatty change within the tubular epithelium but a characteristic interstitial inflammation with infiltration of lymphocytes, plasma cells and eosinophils is the main feature (McMannus 1950) Sulphonamide anuria commonly occurs during the treatment of throat and respiratory infections when the fluid intake has been inadequate Oliguria with the passage of blood and perhaps of crystals, precedes complete cessation of function and the disease then runs the course already described

Symmetrical cortical necrosis—The type of symmetrical cortical necrosis commonly associated with pregnancy has been described in children after severe infectious illnesses The area of necrosis corresponds so closely to that rendered ischaemic by the diversion of blood through the juxta medullary zone, that it is reasonable to suppose that these cases result from the prolonged operation of that diversion The clinical picture described has been haematuria and anuria following severe sepsis elsewhere in the body and all known cases have run a fatal course (Campbell and Henderson 1949)

Calculus anuria—Calculus anuria resulting from the obstruction of both kidneys, obstruction of a solitary kidney or obstruction of one kidney with the reflex failure of the other is exceptionally rare and need not be described in detail here Provided the possibility is considered the diagnosis will not be in doubt (Joly 1934)

Acute suppurative pyelo-nephritis (the surgical kidney)—This occasionally complicates surgical interference in lower urinary obstruction or may rarely be of spontaneous origin The excretion of a diminishing quantity of dilute urine is characteristic and unless the infection is rapidly controlled by antibiotics the combination of renal failure and infection proves rapidly fatal

Acute glomerulo-nephritis—This may be of such severity as to cause anuria and a dangerous uraemia In contrast with the diseases previously mentioned the glomerulus is primarily at fault and any urine which is passed has been concentrated during passage through the tubules and is of normal or high specific gravity (Fishberg, 1939) Accompanying hypertension and oedema of characteristic distribution may assist in the diagnosis

Thrombosis of the renal vein—Thrombosis of the renal vein with consequent cessation of renal function is not very uncommon in association with neonatal gastro-enteritis sepsis and dehydration In most reported cases there has been a sudden onset of pain and shock haematuria and a rapid enlargement of both kidneys Oliguria or anuria follows and is fatal within a few days Sandblom (1948) and Campbell and Matthews (1942) report unilateral cases of venous thrombosis in neonates who were not suffering from any other serious disease Haematuria, pain and enlargement of the kidney should give rise to the suspicion of this condition retrograde pyelography shows no filling of the calyces but a wide diffusion of the dye throughout the renal area Nephrectomy may be curative if performed early in a unilateral case, but we have not yet encountered a suitable one (Such a case has been described by Parry 1951)

Pre-renal uraemia—Oliguria and a rise of blood urea, even as high as 100 milligrams per cent, may result from dehydration, from alkalosis and from other extrarenal factors not clearly understood. In fatal cases of this type no very obvious changes are found within the kidney, though the oliguria may well have resulted from a moderate diversion of the blood away from the cortical glomeruli The small amount of urine passed has characteristically a high urea but a low chloride content (Zondek, 1948), and has a normal specific gravity Dehydration and alkalosis from severe vomiting are particularly likely to occur in the young, and this type of uraemia must be distinguished from true renal disease, but " pre-renal " factors may well complicate chronic lesions of the kidney and be responsible for exacerbations of chronic renal failure

Diagnosis

In many cases the cause of acute renal failure is sufficiently established by the history of the condition, though the rapidly fatal diseases such as venous thrombosis and cortical necrosis may well be missed A straight skiagram to exclude calculus anuria is essential, and in all cases in which a sulphonamide has been administered ureteric catheters must be passed to release any obstruction to the pelvis or ureter Evidence of dehydration or other factors likely to be responsible for pre-renal uraemia must be sought, and the concentration of any urine passed must be estimated

Treatment

The guiding principle of treatment must be to preserve the child's water and electrolyte balance until the kidney recovers its function, and then to make up promptly the abnormal losses involved in the diuresis High blood urea levels are of much less danger than pulmonary oedema Diuretics have no action upon necrotic tubular cells

The daily extrarenal fluid loss of an adult is reckoned as 1,000 millilitres and in the child this will be appropriately scaled down No more fluid than can be lost through the skin, lungs or intestines must be allowed to enter the body The toxic effects of the break-down products of protein metabolism must be minimized by eliminating protein from the diet and by sparing the endogenous protein by a high carbohydrate intake Such a diet may be so nauseating that it needs to be given through a stomach tube, and Bull, Joekes and Lowe (1949) found that by employing this route in adults the following formula could be administered and constituted an unusually successful line of treatment Amounts can be adjusted for use in children

Glucose	400 grammes	per 24 hours
Peanut oil	100 grammes	
Acacia *q s*	to emulsify	
Water to	1,000 millilitres	

Fluid and electrolytes may be lost in vomit, which can be strained and returned to the stomach or rectum Other attempts to correct chloride loss or disturbances of the *p*H should only be made during the period of anuria when the imbalance is extreme, but once the diuresis starts it is essential to replace the water quantitatively and chloride loss may then necessitate intravenous therapy

These general methods of preventing over hydration and minimizing the rise of d urea may be applied to all cases of established anuria in which a pre renal e can be eliminated Certain specific measures however are required for ific diseases In sulphonamide oliguria it is essential to know whether or not tals are present and whether they are constituting an obstruction to the extra l passages Ureteric catheters should be passed and the ureters washed out uld this measure reveal the presence of obstructing crystals particularly when ipper respiratory infection has caused the child to drink very little fluid intake t be increased and alkali administered but with caution Pyelostomy is isionally required to overcome the obstruction Where crystal blockage does appear to be the factor responsible for anuria the routine described above t be instituted In anuria following mismatched transfusions, heavy doses of lis may be effective Calculus anuria constitutes an obvious indication for ;ery

Decapsulation of the kidney in almost any type of acute failure has occasionally i followed by a diuresis, an effect attributed to the release of tension but it is cult to know how far the diuresis in such cases has been due to coincidental ular recovery Infiltration of the renal pedicle with procaine and intravenous caine injections have been reported as of value in the sulphonamide cases ugh their mode of action is not clear (Chauvin, 1949 Friis, 1949) Splanchnic ipathetic block performed in the hope of restoring blood to the renal cortex the lower nephron nephroses is seldom helpful Diuretics should not be iloyed in established anuria, but in doubtful cases of oliguria isotonic sodium ihate (4 285 per cent) given intravenously is comparatively innocuous and y be helpful

When the patient is already water logged because of the misdirected administra i of large quantities of fluid given in the hope of bursting the dam itoneal irrigation or the use of the artificial kidney may be justifiable Neither these methods is free from serious complications and they should not be ployed unless pulmonary oedema is an imminent danger Intestinal and gastric age have been attempted in the hope of eliminating the risks but they are not so ective as peritoneal irrigation which remains the most easily applicable form of lysis Catheters are inserted through small wounds, one into a flank and the ier into the pelvic peritoneum An isotonic fluid Tyrode or Hartmann is run o the upper catheter and drawn off through the lower The amount of water sorbed from the irrigating fluid can be regulated by varying the concentration glucose and throughout the process a constant check must be made upon the od chemistry

For the discussions of peritoneal dialysis the reader is referred to Kolff and rk (1944) Swan and Gordon (1949) and Fields *et alii* (1949) The following case istrates the use of this method

I D Male. Aged 2 years 2 months.

Was admitted in June 1947 from another hospital, where he had been treated for gastro-enteritis with sulphasuccidine (2 grammes 4 hourly for 3 days) No urine had been passed for 3 days before admission despite intravenous therapy and the admin stration of sodium sulphate The blood urea was 316 milligrams per 100 ml. Cystoscopy was performed and the right ureter catheterized Four millilitres of blood-stained

urine were recovered from this catheter but no sulphonamide crystals On the following day decapsulation of the right kidney was performed, the renal substance being tense and bulging but the pelvis and ureter unobstructed Only 2 ounces of urine were passed during the 2 days following this operation, and oedema appeared in the genitalia and legs Peritoneal dialysis was then started using full strength Hartmann's solution and 5 per cent glucose Fig 6 shows the effect on the blood urea, which fell rapidly during

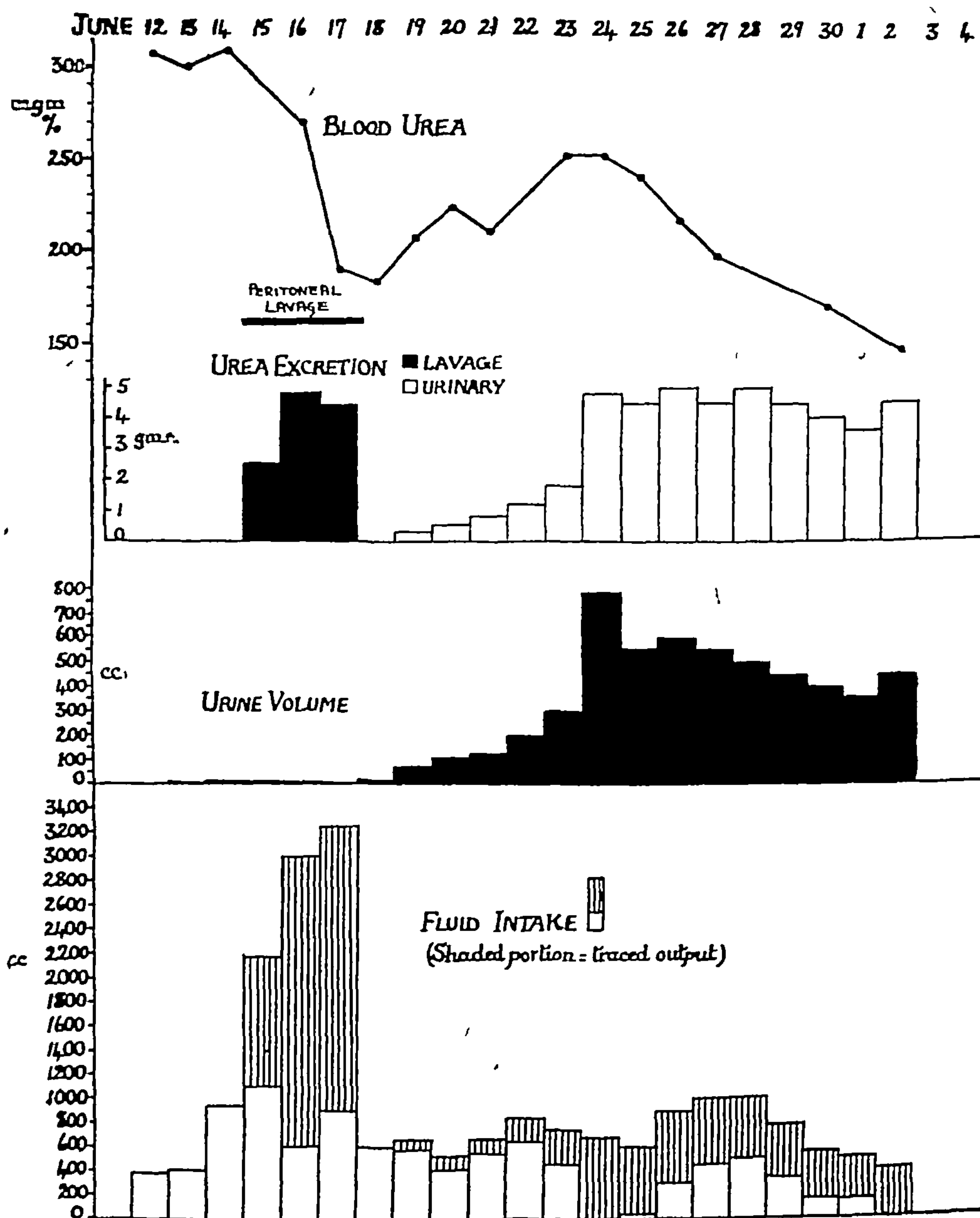


FIG 6 —Chart showing effect of peritoneal dialysis in acute renal failure in a boy of 2 years 2 months

the period of irrigation, but it will also be seen that a proportion of the fluid run in could not be withdrawn and the oedema became worse After 3 days the irrigation was discontinued and a slowly increasing volume of dilute urine was passed The blood urea, however never returned to normal and although for some time the child's general

condition appeared to be good, he was re-admitted 3 months later with uraemia to which he succumbed A post-mortem examination of th- kidneys showed advanced damage to the parenchyma and some calcification.

Chronic renal failure

A wide variety of diseases in children may lead to renal failure of gradual onset with a progressive destruction of the renal parenchymal tissue Among these diseases are bilateral hydro-nephrosis chronic pyelo-nephritis alone or com plicating obstruction or stone congenital hypoplasia of the kidney chronic glomerular nephritis periarteritis nodosa renal tuberculosis and polycystic disease Where urinary obstruction is not, or has ceased to be, a factor almost any of these conditions may progress until the kidney is a scarred and shrunken organ *which is incapable of recovery and in which all stigmata of the original disease* have been lost. In the obstructive cases however and in the cases of pyelo-nephritis in which active inflammation rather than scarring is responsible for the depression in renal function some recovery is possible

Rare cases are encountered in which there is apparently a specific failure of certain functions of the renal tubular cells Such failure may be *congenital* as in Fanconi s disease a vitamin D resistant type of rickets in which there is inadequate tubular re absorption of glucose and amino acid including cystine or *acquired* as in the nephro-calcinosis associated with hyperchloraemic acidosis in infants This latter condition is discussed in Chapter 11 In the great majority of diseases however all aspects of tubular function are depressed at the same time

Since obstruction and infection hydro-nephrosis and pyelo-nephritis are the chief concern of the urinary surgeon it may be well to mention briefly their pathology In both conditions the tubules are first affected and functional tests at first show a depression of tubular rather than glomerular function (Raaschou, 1948) The renal parenchyma is thinned out in a hydro-nephrosis and there is a moderate tubular atrophy but microscopical changes are never very marked where the condition is bilateral By contrast in the unilateral cases in which overall renal failure is not an important factor the tubules having been at first dilated, become shrunken and atrophic while the intact glomeruli remain for some time a striking feature of the histological picture Later however the glomeruli become hyalinized as a result of disuse In uncomplicated pyelo-nephritis which in its bilateral form is perhaps one of the commonest causes of chronic renal failure the kidneys are small and scarred but on cross section it can be seen that areas of fibrosis alternate with areas of surviving tissue The tubules are dilated and their epithelium atrophic many contain colloid casts giving them the appearance of thyroid tissue The glomeruli are not damaged in the early stages but capsular adhesions, periglomerular fibrosis and hyalinization are later evident. Interstitial fibrosis and leucocytic infiltration are prominent features and the blood vessels show a variable degree of pathology hyperplastic arteriolosclerosis being associated with hyper tension (*vide infra*) Chronic pyelo-nephritis with renal failure may be associated with well recognized chronic or recurrent urinary infection with urinary calculi with congenital abnormalities of the urinary passages and with hydro-nephrosis but quite often a definite history of the infective onset is lacking, and organisms in the urine are hard to find In this latter form pyelo-nephritis is particularly

difficult to distinguish from chronic glomerulo-nephritis, but in neither case is treatment likely to be successful

Each member of this group of diseases, characterized by chronic renal failure, has its peculiar features, but variations in the symptoms attributable to failure itself depend upon the rapidity of the onset rather than on the individual lesion and upon the presence or absence of hypertension

The failure of renal function naturally involves a rise in blood urea more or less proportional to the degree of damage, but in very chronic cases high figures may be found in ambulant patients, while with a more rapid onset the child may be moribund with a blood urea of no more than 100 The term "uraemic" may justifiably be applied to all cases in which the blood urea is raised, but it is commonly limited to those with severe and often terminal symptoms

The kidney loses its capacity to form a concentrated urine and many of these children have marked polyuria and thirst The urine is pale and the specific gravity may not normally exceed 1 002–1·004, though higher figures (1 006–1·010) may be achieved by dehydration The cause of the polyuria is not very obvious but it is probably related to the small number of functioning nephrons, since Bradford (1899) found that by removing a large proportion of the renal tissue of experimental animals he could induce a severe polyuria The excessive amount of urine formed may itself be responsible for the nocturnal enuresis or frequency which first draws attention to the renal disease Whatever the cause of failure, the urine is likely to contain a small amount of albumin and casts may be present, particularly in the glomerulo-nephritics

A failure of growth and deformities of the bones are common whenever the disease has started early and run a very prolonged course The children are dwarfed, wasted and there may be actual infantilism with a failure of development of the secondary sexual organs The mechanism of this interference of growth is not precisely known but it may be reversible and the dramatic metamorphosis, which may follow simple drainage in the obstructive cases, is one of the most encouraging features of children's urology. The bone changes are commonly spoken of as "renal rickets" and are in general very similar to those seen in hypovitaminosis D rickets, though much later in onset They occur most often in the slowly progressive renal failure associated with bilateral renal hypoplasia Genu valgum is the most common deformity but the wrists will show radiologically the characteristic broadened and "woolly" metaphysis (For the discussion of the bone lesions the reader is referred to Parsons, 1927) Bone changes are accompanied by hyperphosphatemia and by acidosis In all types of renal failure, particularly in infants, there is a tendency towards acidosis due to the poor excretion of phosphates and of other acid radicals, and this acidosis alone may be responsible for osteodystrophy The raised serum phosphorus, however, which may be as high as 10 milligrams per cent, results in depression of the blood calcium and hypertrophy of the parathyroid glands with specific bone changes

Anaemia of the hypochromic type is common in renal failure and there is frequently a yellow, or greyish-yellow pigmentation of the skin Oedema is very seldom seen in these diseases in contrast to the cases of acute glomerulo-nephritis and of nephrosis When it occurs it is usually terminal, the result of cardiac failure or misdirected treatment

The hypertension which may or may not accompany renal failure is discussed in a later section

In the terminal stages of a progressive renal failure or in the earlier stages of a more rapidly destructive lesion true uraemic symptoms appear headaches nausea and vomiting, diarrhoea coma and convulsions, stertorous or acidotic respiration The polyuria may have resulted in dehydration which is aggravated by the fluid loss in diarrhoea and vomiting Disturbances of the electrolyte balance are then likely particularly depression of the serum chloride The usual uraemic tendency towards acidosis may be reversed if vomiting is severe and particularly if urinary symptoms have been treated with an unnecessary amount of potassium citrate, alkalosis is a danger Potassium intoxication has been mentioned as a cause of heart failure in uraemic cases

Management

In general the symptoms of renal failure are such as to bring the patient to the physician upon whom falls the burden of watching the terminal stages of an incurable disease Once chronic pyelo-nephritis or glomerulo nephritis has reached the stage of a small contracted kidney the most that can be done is to help the child through the lesser crises which precede the final collapse In the potentially reversible cases, however particularly with obstruction to the urinary passages no time must be lost in securing adequate drainage Diagnosis of the obstructive lesion is frequently obvious because of the enlargement of both kidneys and the disorders of micturition Straight skiagrams are important in eliminating the possibility of renal calculi but pyelograms are of little assistance since the damaged kidneys will not excrete diodone in adequate concentration while retrograde catheterization or indeed any method of urological investigation, is apt to precipitate a dangerous exacerbation The severe infections will be diagnosed from the urinary findings and of the other causes of bilateral enlargement of the kidney only polycystic disease is likely to cause confusion It is however very rare in this age group It must not be forgotten that one kidney may be absent or the two may be fused or ectopic

The methods of drainage in obstructive lesions are described in Chapter 8 with regard to correction of the biochemical disorder it must be emphasized that dehydration is usually the most urgent problem and that fluid is far better restored to the body through the mouth than through the veins In serious cases blood urea tests should be supplemented by estimations of the serum chlorides and of the plasma CO_2 Severe loss of chloride usually the result of vomiting, is an indication for intravenous therapy Half normal saline (0·45 per cent) made up to isotonicity with glucose is preferable to physiological saline and as soon as the salt deficiency is rectified only glucose (5 per cent in water) should be given Acidosis and alkalosis seldom need specific correction in the older child with urinary obstruction but blood transfusion may be urgently required at the time of the establishment of urinary drainage

In infants in their first year of life, biochemical disturbances are particularly dangerous and especial care is required in the administration of intravenous fluids. The value of a ward sister with experience of these disorders in infancy and of a biochemist with sufficient clinical contact to enable him to adjust the nature

and amounts of parenteral fluids can scarcely be over-estimated, and all young infants suffering from serious urinary conditions should, if possible, be transferred to an institution providing these facilities The normal fluid requirements of an infant are approximately 150 millilitres per kilogram of body-weight per 24 hours, though a little more may be necessary in cases of polyuria Quite often replacement of the required amount of fluid by mouth is impossible, and subcutaneous or intravenous infusions have to be given Over-dosage with saline, even when half-normal solutions are used, is always a danger and serum values should be estimated repeatedly Acidosis may be corrected by the use of Hartmann's sodium lactate solution, either by mouth or by vein

Hypertension

Severe hypertension is rare in the young but in those instances which are not explained by coarctation of the aorta, hypothalamic tumour, hyperthyroidism, acrodinia or some systemic mineral poison, urological examination becomes of the utmost importance Essential hypertension has indeed been described in children (Court, 1941, Sobel, 1941) but should only be diagnosed after a careful exclusion of other causes

The normal blood pressure in children may be seen from Table 1 in Chapter 2 All observers agree that a narrow sphygmomanometer cuff gives higher readings than a broad one, though the reason for this is obscure The cuff used should cover at least half the length of the upper arm, the child must be at rest when the reading is made and accustomed to his surroundings Single observations in the out-patient department are not adequate

The symptoms and signs of hypertension in the child do not differ from those seen in the adult and need not be described here The function of the urologist in these cases is to investigate the possibility of unilateral renal disease being responsible for the high blood pressure or to localize a possible phaeochromocytoma

Renal hypertension

The pioneer work of Goldblatt (1947) and of Wilson and Byrom (1939) has made it clear that hypertension may be produced in the experimental animal by rendering one kidney ischaemic Some humoral factor appears to be responsible and the presence of a normal contra-lateral kidney tends to prevent the appearance of hypertension High blood pressure itself, however, may be responsible for vascular changes in the normal kidney, which will perpetuate the condition after the original cause has been removed (For review of experimental work see Goldblatt 1947)

A great deal of evidence suggests that the human kidney reacts to ischaemia in a way similar to that found in the experimental animal, but in pathological conditions the interference with the blood supply is seldom so simple as a constriction of the renal artery All diseases which cause damage to the renal parenchyma have on occasions been associated with hypertension, but it is least common in the simple uninfected obstructions and most common where scarring and fibrosis is maximal

Atrophic pyelo-nephritis and congenital hypoplasia of the kidney are the most important diseases to be considered Not all cases of chronic pyelo-nephritis

have hypertension even when the disease is bilateral Kimmel (1942) for instance found high blood pressure in only 10 per cent Weiss and Parker (1940) believed that there was a significant relationship between the incidence of hypertension and the degree of vascular change (hyperplastic arteriolosclerosis and endarteritis obliterans) but other observers have not found so clear-cut an association and point out that these changes may be the result rather than the cause of the hypertension

It seems unlikely that the congenitally small but histologically normal kidney can be the cause of hypertension The scarred and contracted kidney which is so small that an origin in congenital hypoplasia is suspected is frequently found to be responsible for the raising of the blood pressure scarring in these cases may have resulted from superadded pyelo-nephritic change but apparently sometimes takes place without infection Ectopic and double kidneys are unusually liable to hypoplasia and may be important in this connection

Braasch *et alii* (1940) have drawn attention to the incidence of hypertension following conservative operations upon the kidney presumably the result of operative trauma and infection and in undertaking any such procedure one should bear in mind this possibility Particularly is this true when dealing with a unilateral case of hydro-nephrosis where the preservation of a partially defective kidney might lead to a far more serious disease and where there is well marked fibrosis in the kidney nephrectomy should be performed Follow up readings of the blood pressure are essential after all conservative renal surgery

Sterile hydro-nephrosis is seldom associated with hypertension though Bell (1946) records a case of high blood pressure in a child with uninfected lower urinary obstruction and bilateral hydro-nephrosis

Polycystic disease is rare in children but may occasionally be a cause of hypertension (Bothe 1939) Renal tuberculosis despite the frequently advanced scarring of the renal substance is exceptionally seldom responsible for this condition Of the purely medical diseases of the kidney chronic glomerulo-nephritis and periarteritis nodosa are well known causes

Several authors (for example Daniel 1939) have noticed a moderate raising of blood pressure associated with Wilms tumours The mechanism in these cases is presumably different from that found in the scarred kidney In our cases high blood pressure has not been a significant feature

Although every kidney responsible for hypertension is to some extent damaged from the point of view of its ability to secrete urine, there is no direct relationship between the degree of renal failure and the raising of the blood pressure. Thus of 20 renal dwarfs recorded by Ellis and Evans (1933) in whom the parenchymal damage was extreme and of long standing, there were only 2 with hypertension

From the surgical point of view it is the cases of unilateral disease which are of interest. if the damaged kidney can be removed before the hypertension has had time to produce secondary changes in the contra lateral kidney there is a chance of permanent cure The selection of cases suitable for surgery depends very largely upon the intravenous pyelogram Where one kidney is found to be normal or hypertrophied and the other has diminished function or complete absence of secretion the case should be given further consideration Retrograde pyelography is performed in order to exclude the possibility of complete renal agenesis,

and the outline of the pelvis of the damaged kidney gives important clues as to the nature of the disease Frequently the calyces are narrow and bunched together, while the pelvis is a little dilated Such an appearance may be produced either by atrophic pyelo-nephritis or by congenital hypoplasia of the kidney, and in either case, provided the hypertension is of comparatively recent origin, a nephrectomy is indicated In other instances there may be some clear-cut urological indication for operating upon the kidney, as in the case of renal calculus, and in these nephrectomy is to be preferred to conservative operations provided the contralateral kidney is normal

Where a unilateral renal disease is found to be responsible for hypertension and nephrectomy performed, the blood pressure falls immediately to a more or less normal level Unfortunately this gratifying fall is not always maintained, and the pressure slowly rises again during the weeks following the operation and appears to stabilize at a level about half-way between the normal and the pre-operative reading The child may not be cured but the symptoms are satisfactorily alleviated and the progress of the disease appears to be arrested

The following cases are examples of renal hypertension

D L Female Aged 11 years

This case was referred by Dr Paterson for urinary investigation She had a blood pressure of 218/174 with headaches, dimness of vision and retinal changes The urine contained pus cells The intravenous pyelogram showed poor function on both sides but the retrograde outlined normal, though somewhat spastic, renal pelves and ureters Since the lesion appeared to be bilateral no surgical treatment was undertaken and there was a slow deterioration in the general condition of the child She died 1 year later The blood urea did not rise above 64 mg per 100 ml until immediately before the end

N G Female Aged 11 years

This case was referred by Dr Paterson and was later under the care of Dr Sheldon She complained of headaches, puffiness of the ankles and enuresis The blood pressure was 205/130, retinal changes were present The urine was sterile, the blood urea 38 The intravenous pyelogram showed a large right kidney with a double pelvis, and a small and poorly functioning left kidney A left nephrectomy was performed, the specimen showing a hypoplastic organ containing areas of normal parenchyma and areas of gross fibrosis Arteriolar thickening and hyalinization were noted microscopically Immediately after operation the blood pressure was 150/120 14 days later it was 186/136 One year later it had risen to 200/145, and the prognosis was considered poor The enuresis remained unchanged throughout the period of treatment

C H Male Aged $5\frac{1}{2}$ years

This case was referred by Dr Paterson with attacks of pain in the left loin The urine was infected with *B coli*, the blood urea was 41 mg per 100 ml, and the blood pressure 150/105 The intravenous pyelogram showed a normal right kidney and a poorly functioning one on the left, which was shown by retrograde to be hydronephrotic A conservative operation (division of aberrant vessels) was performed on the left kidney and the infection cleared with sulphonamides After the operation there was no recurrence of pain but the blood pressure remained unchanged On re-admission 7 months later it was 165/120 and a nephrectomy was performed At the end of this operation the blood pressure was 135/85 and 18 months later 110/70

P T. Female Aged 13 years

Referred for urological investigation by *Professor Moncrieff* There were no urinary

symptoms the child was suffering from headaches and vomiting and was found to have a blood pressure of 255/180 Papilloedema was present. Blood urea was 30 milligrams per cent Intravenous pyelogram showed a normal right kidney but very little secretion on the left side At cystoscopy there proved to be 2 left ureteric orifices, both of which were catheterized The upper pelvis was slightly dilated with a blunted calyx and the calyces of the lower pelvis were also small The urine contained a few pus cells but was sterile on culture Left nephrectomy was performed. The specimen showed a hypoplastic double kidney (*see* Fig. 7) Fourteen days after the operation the blood pressure had fallen to 180/150



FIG 7—Hypertension Case P T Operation specimen Double left kidney showing chronic pyelo-nephritis.

K H Male Aged 4 years.

First admitted under Dr Lightwood with frequency enuresis and attacks of abdominal pain he was found to have a blood pressure of 210/135 During the course of the following year the blood pressure varied between this figure and 150/100 and he had attacks lasting a few days of frequency thirst and irritability The urine was sterile but always contained albumin The retinae remained normal Intravenous pyelography showed good function in both kidneys, but the right pelvis was large and normally formed whereas the left was small and narrow (*see* Fig. 8) Retrograde pyelograms did not give any further information In view of the periodicity of the attacks, a phaeochromocytoma was considered as a possible cause, and the adrenal areas were investigated by radiography after air insufflation No tumour was demonstrated. Dibenamine (5 milligrams per kilogram) was given intravenously without any significant effect upon the blood pressure. Laparotomy revealed no sign of a phaeochromocytoma either in the adrenals or elsewhere in the abdomen and both kidneys appeared to be of normal consistence although the left one was very small The blood pressure then tended to stabilize at the higher level (270/180) and a left nephrectomy was performed

The excised kidney was small but macroscopically normal Histologically there some evidence of pyelo-nephritis In the 2 months following the operation the pres fell to 150/100, and 6 months later the boy is symptom-free, extremely well and blood pressure is 120/80



FIG 8 —Excretion pyelogram boy with se hypertension, aged 4 years Note good func on both sides and narrow left pelvis with m calyces obliterated

Phaeochromocytoma (paraganglionoma)

Although it has no connection with disorders of renal physiology it is conveni to discuss here another cause of hypertension in children—the tumours of chromaffin tissue

The phaeochromocytomas are adrenaline-secreting tumours which arise from medulla of the supra-renal gland or in other elements of the sympathetic cha They are histologically benign but their secretory output induces deadly hyp tensive cardiovascular disease This takes the form of a steady and progress elevation of the blood pressure associated characteristically, but not invarial with paroxysmal exacerbations of hypertension during which the child suff from severe headaches, palpitations, excessive sweating, polydypsia and polyu The increased urinary output may be evidenced by frequency and enuresis T tumours are usually small, though at least one (Snyder and Vick, 1947) has be palpable in the pararenal area In childhood, multiplicity of the tumours has be a feature of several cases (Cahill and Aranow, 1949)

Diagnosis

Diagnosis is likely to be a matter of some difficulty Paroxysmal attacks, if th are a feature, are very suggestive, but even in their absence phaeochromocyto is always a possibility The demonstration of an excess of adrenaline in the blo has not proved to be a satisfactory investigation, and the practice suggested Roth and Kvale (1945) of provoking a typical attack by the injection of histam

is not without dangers The use of substances which annul the effect of adrenaline has proved to be a more successful approach and in particular dibenamine has been used with great effect This investigation was described by Spear and Griswold (1948) who advised the intravenous injection of 7 milligrams dibenamine hydrochloride per kilogram of body weight In cases of phaeochromocytoma the blood pressure may fall to normal for a period as long as 24 hours

A pyelogram in these cases not only assures the absence of any renal disease but may indicate the presence of a supra renal tumour by displacement of the renal pelvis. The injection of oxygen into the perirenal space is also a valuable means of demonstrating the tumour The child is anaesthetized and a needle connected up with a manometer and an oxygen supply is introduced into the angle between the last rib and the erector spinae and thrust gently forwards and medially Once the perirenal space is entered oxygen flows in freely at a comparatively low pressure and exposures are then made

These investigations may localize a phaeochromocytoma when it is situated in the supra renal gland but when it is located elsewhere in connection with the abdominal sympathetic it can only be found by exploratory laparotomy Also tumours may rarely occur in the thorax or the neck

Treatment

Treatment of these tumours is clearly operative but the risks involved either from an excessive release of adrenaline caused by operative manipulation or from a dangerous fall in the blood pressure when the adrenaline supply is cut off must not be underestimated Successful removal has been accomplished in a child and with earlier diagnosis and with the better operative management to be expected from a fuller understanding of the condition satisfactory results may be anticipated in the future.

Note on glomerulo-tubular nephritis: the surgical aspect

A textbook primarily surgical is not the place for a dissertation on the medical diseases of the kidney but there are certain aspects of glomerulo-tubular nephritis which have a bearing on the urological lesions of childhood

It has been shown (Davson and Platt, 1949) that the Ellis classification (1942) of nephritis has stood the test of time with very few exceptional cases

Type I nephritis is characterized by an initial acute illness with haematuria, hypertension and oedema Over 80 per cent (Davson and Platt, 1949 Ellis, 1942) recover and do not relapse Some others pass into a rapidly progressive phase with continuing hypertension and oedema with early uraemic death Others run a chronic progressive course lasting years and are sometimes quite symptom free for a long period before diminution of renal function produces a return of symptoms. The acute onset phase may be overlooked or missed having been masked by an acute infective episode such as tonsillitis, which so often precedes this type of nephritis

The Type I nephritis, becoming slowly progressive from early life produces a shrunken kidney with no features to differentiate it from one of chronic atrophic pyelo-nephritis which itself may be superimposed upon congenital renal hypoplasia

Type II nephritis is characterized by minimal (often microscopic only) haematuria, disproportionately gross albuminuria and oedema much more marked in duration and degree This type is rarely confused with any surgical lesion

Acute focal nephritis with haematuria but no oedema or hypertension, often occurring during an acute infection such as tonsillitis, appendicitis or osteitis, is an entity which is considered worthy of separation from the Ellis types (Davson and Platt, 1949) Recovery is the rule without progress to a chronic stage, but often with recurrences in subsequent infective states

The surgeon must then bear in mind particularly the Type I nephritis and acute focal nephritis, and periarteritis nodosa, as causes of haematuria which may be very hard to distinguish Such lesions may occur whilst a urinary infection is present, and precipitate a renal failure which would be unexpected purely from the degree of the infection Haematuria occurring during treatment of acute throat infections by sulphonamide drugs may be due, in fact, to such primary nephron disease and not to damage by the drug itself

CHAPTER 5

INFECTIONS OF THE URINARY TRACT

Pathology and clinical features

THE prevention and control of infection plays so large a part in children s urology that some general review of the aetiology pathology and treatment is necessary Acute infections of the normal urinary tract often short lived produce a characteristic clinical picture still commonly termed pyelitis despite the fact that there is no real evidence in most of upper tract involvement. In true pyelitis pathological evidence shows that there is always an inflammatory process within the interstitial tissue of the kidney whenever the pelvis is involved, that is pyelonephritis. The term *pyuria* is a simple description of the common finding, which does not commit the clinician to a definite statement of the site of the inflammation nor does it exclude the possibility that a more serious abnormality or disease may be present. The management of the acute transient infections remains in the hands of the physicians and the urologist is concerned chiefly with those cases in which the condition is progressive persistent or recurrent

Bacteriology

The organisms most commonly found in the urine belong to the *B coli* group though there is perhaps more variability than was at one time suspected and other gram negative bacilli as *Aerobacter Proteus vulgaris* and *Pseudomonas pyocyaneus* may be present, as well as the gram positive *cocci Staph albus* occasionally *Staph. aureus* and *Streptococci* usually of the *faecalis* type Mixed infections are not uncommon and the organism may change during the course of treatment, a type susceptible to chemotherapy being too often replaced by an insusceptible one These variations however occur chiefly where there is a complicating abnormality and in the transient infections *B coli* predominates

Source of infection

The gram negative bacilli and *Strep faecalis* are clearly of bowel origin and the intestine is the factory to which due attention should be paid in the treatment The intestinal disturbances so common in early life and especially in infancy must determine striking variations in the bacterial content and its virulence, an aetiological factor of great importance in urinary infections Tonsillar infection and otitis media have also some significance in our experience

Route of infection

The route by which the organisms reach the urinary tract is a matter of controversy there can be no doubt that a blood-stream spread direct to the kidney does often occur but from the facts of common observation it seems manifest to us that in the majority the organisms first reach the bladder *via* the urethra and that

involvement of the upper tract is by no means so common as is generally supposed During the "diaper" age, in which the disease is so frequently seen, there is ample opportunity for contamination of the urethra, which is a comparatively short structure in both sexes at this time later in childhood, infection is considerably more common in girls, again because of the opportunities of contamination and the short ascent The simple lodgement of the organisms within the bladder is not of course sufficient to produce a cystitis, and a subsidiary cause is commonly found, such as depression of the general health and lowered resistance due to defective diet, or exposure to cold and damp, increase in the virulence of the intestinal organisms or urinary stasis The great majority of children cystoscoped during an attack of "pyelitis" show at least a basal cystitis, and this alone may be responsible for the symptoms the uretero-vesical buffer mechanism remains effective and the upper tract never becomes involved In the serious infections, however, the oedema round the ureteric orifice renders the valve incompetent, and reflux of vesical contents carries the organisms upstream The resulting ureteritis spreads the dysfunction so that the pelvis and calyces are infected and a true pyelitis and pyelo-nephritis ensue Any pre-existing anomaly, congenital or acquired, will obviously expedite this process

Although the lymphatics round the ureter and along the fascia of the posterior abdominal wall may possibly play a part in the ascent from bladder to kidney, we are inclined to regard the lumen of the disorganized ureter as the most important channel

Pathology

Examination of post-mortem material from infants who have died with pyuria, almost always accompanied by gastro-enteritis, meningitis or otitis media, has shown a predominance of changes in the kidney (Chown, 1927, Wilson and Schloss, 1929) It seems clear that in such cases the renal infection is blood-borne, but we believe that this does not give an accurate indication of the common onset of the disease, though the lesions in the kidney are certainly those which endanger life during the acute attack and may be responsible for symptoms later on

During the attacks of acute pyelo-nephritis there is a generalized inflammation of the kidney The renal pelvis is reddened and oedematous Within the renal substance there is a pronounced interstitial leucocytic infiltration, which in more severe cases may go on to suppuration, the minute abscesses showing as radiate streaks upon the cut surface of the kidney On rare occasions this suppuration proceeds unchecked with destruction of all renal elements, and abscesses may rupture into the perinephric space Commonly, however, healing occurs quite quickly, leaving little dimpled scars on the surface of the kidney and areas of fibrosis throughout the parenchyma

In chronic pyelo-nephritis, in recurrent acute attacks, or even in the healed disease, there may be a progressive destruction of the nephrons This is not a uniform process taking place throughout the kidney, but scarred areas alternate with normal or with hypertrophied parenchymal tissue The whole kidney is contracted as a result of the scarring and on the cut surface white and grey or yellow streaks of fibrosis are evident When the disease has occurred very early in life, the fibrotic kidney is naturally unable to grow and appears very small in

relation to the size of the body so that a congenital hypoplasia may be suspected indeed the differential diagnosis between atrophic pyelo-nephritis and congenital hypoplasia is frequently difficult, if not impossible

Histologically interstitial fibrosis is a prominent feature the tubules are dilated and lined by an atrophic epithelium they contain colloid casts and have an appearance in cross section closely similar to thyroid tissue The glomeruli remain intact for longer than the tubules but ultimately capsular adhesions periglomerular fibrosis and vascular changes put them out of action Endarteritis obliterans and hypoplastic arteriolosclerosis are commonly found particularly in cases associated with hypertension

Pyelo-nephritis may and frequently does complicate hydro-nephrosis and renal lithiasis and the gross appearance of the kidney may be considerably altered by the primary disease The microscopical changes within the parenchyma and their physiological effects do not differ materially from those already described A renal calculus may cause severe ulceration of the pelvic mucosa and scarring of the renal substance is then apt to affect chiefly the pyramids whose apices have been damaged

The pelvis and upper ureter are usually somewhat dilated in the early stages of uncomplicated pyelo-nephritis, but with progressive contraction of the renal substance the calyces are drawn closer together and narrowed

Clinical

In infants acute urinary infections give rise to very few urinary symptoms high fever or sudden onset with vomiting, diarrhoea and perhaps convulsions constitute a characteristic clinical picture which may cause confusion with meningitis or pneumonia Early examination of the urine, which is slightly turbid and contains pus cells on microscopy reveals the true cause of the symptoms

In older children there is less constitutional disturbance and though fever and vomiting may be the chief complaints there is almost always frequency and dysuria Abdominal pain rather poorly localized and not very severe is a feature of some cases and may be mistaken for appendicitis in a few cases there is a sudden onset of severe pain in the loin with rigors as in the pyelitis of adult life The febrile stage of the acute transient infections does not last more than a few days and less if treatment is started immediately The pyuria may continue a little longer

At times however the fever does not subside and it becomes clear that a suppurative process is occurring in the kidneys, one or both of which will be palpably enlarged and tender Such an enlargement may be due to a suppurative pyelo-nephritis, which is apt to lead on to a perinephric abscess,* but is more likely to indicate that infection has supervened upon a previous abnormality of the kidney A pyo-nephrosis* commonly results from an infection complicating a congenital hydro-nephrosis due to obstruction at the pelvi ureteric junction but a stone is occasionally responsible for the obstructive element. Where infection supervenes upon a long standing lower urinary obstruction it may precipitate acute retention or an acute exacerbation of renal failure, which is apt to overwhelm the child before treatment can be effective

In many cases seen by the urologist, the fever and constitutional disturbance

For further description see end of this chapter

have subsided as a result of treatment but the urine remains infected This observation calls for a thorough investigation of the whole urinary tract, and will frequently bring to light some anomaly such as hydro-nephrosis, megaureter, or duplication of the ureter, which requires surgical treatment

Recurrent acute or subacute attacks of urinary infection are often observed in children, particularly in girls, and here again investigation will often reveal an anomaly Not infrequently, however, the urinary tract appears entirely normal It is true that in such cases the attacks commonly cease spontaneously after a few years, but it is important to remember that with each additional insult the damage to the renal parenchyma becomes more serious Such a child may go on quite happily until the third or fourth decade of life, only to succumb at that time with pyelo-nephritic contracted kidneys, perhaps complicated by hypertension Since pyelo-nephritis is not infrequently a unilateral disease, it behoves us to be on the look-out for the unilateral contracted kidney which has such a characteristic pyelogram The problems of renal failure and of hypertension are discussed in Chapter 4, but it may be recalled here that the infection has frequently died out by the time the renal damage becomes evident

Treatment

The management of urinary infections will vary a great deal depending upon whether an underlying abnormality is found within the urinary tract, and it is appropriate in this chapter to deal only with the general principles of clinical treatment

Control of the infection is now largely a question of choosing the right antibiotic or chemotherapeutic drug, a matter which is discussed in the succeeding section

Treatment of the underlying obstruction or abnormality may be an emergency measure or a planned approach, and will obviously depend upon the exact diagnosis

An estimate must be made of the degree of renal failure, particularly in the uncomplicated case of pyelo-nephritis For this the intravenous pyelogram may be adequate, but urea clearance or concentration tests may also be helpful

Associated secondary pathological conditions such as anaemia and retardation of growth may require attention and where the disability has been sufficiently severe to interfere with the child's social and educational development, measures may be required to set him back upon the right lines

The management and control of urinary infection

Reference has already been made to the effects of urinary infection on the renal parenchyma, and the urologist is rarely faced with the treatment of a simple straight pyelocystitis More often he is called upon when infection has become established in a damaged urinary tract or when an acute episode has been superimposed on previous chronic infection and urinary obstruction

The establishment of a free flow of urine from the bladder or, in the case of ureteric obstruction, from the kidney is the essential first step The surgeon will have to decide in cases of bladder outlet obstruction whether temporary catheter drainage through the urethra is advisable or whether he should immediately establish supra-pubic drainage or even bilateral nephrostomy It is common to find that where there has been chronic bladder outlet obstruction, especially associated with spina bifida, the urine is so thick with ropy mucus or frank pus

that it is essential to establish continuous catheter drainage with regular wash outs of the bladder Such bladder irrigation may be performed by the usual automatic tidal drainage apparatus, a simple Dukes intermittent flow apparatus, or with a syringe Small girls tolerate the presence of a self retaining urethral catheter fairly well but the calibre of the catheter used on a little boy is usually too small to allow the free flow of debris and mucus so that a supra pubic drainage is more often necessary at this stage It must be emphasized that the major purpose of bladder irrigation is a mechanical one, although antiseptic fluids are used We have been well satisfied with sulphacetamide 1 5 000 and we have found that 2 per cent sodium bicarbonate is a help when the presence of excessive mucus makes irrigation difficult

History of urinary antisepsis

Once free drainage has been established the question of chemotherapy arises and the selection of the appropriate drug requires careful consideration and an assessment of the advantages and disadvantages inherent in each particular substance. The first half of the twentieth century has seen tremendous advances in the control of urinary infection The principal agents available before 1932 were those which acted either by changing the reaction of the urine to one which was unfavourable to the particular organism s survival or by the excretion of some substance which acted as an antiseptic in the urinary tract irrespective of the reaction of the medium in which it would act It is, however, only possible to release formaldehyde in the urine from hexamine given by mouth if the urine is acid Urea-splitting organisms in the urinary tract make it extremely difficult to obtain an acid urine and in children particularly the presence of an infection with *B proteus* may lead to the administration of excessive quantities of acid producing substances such as ammonium chloride The urine will remain alkaline, and extreme acidosis will be produced which may in itself be fatal This is a very real danger and we have stressed earlier the limited control which the infant s kidney has over the variations in its blood acidity

The ketogenic diet which was introduced in 1932 produced one major step forward in attempts to control these infections but was never easy of application in children The beta hydroxybutyric acid thus produced was superseded as a urinary antiseptic by the introduction of mandelic acid in 1936

Prontosil the first of the sulphonamide group of drugs became available in 1937 and opened up a new field of therapy particularly applicable to children

In view of the fact that Prontosil and its successors are bacteriostatic and not bactericidal and that furthermore they are dependent upon the bacterial population present (*see* page 51) the older methods of treatment still have a place, and in 1946 organic combination of hexamine and mandelic acid was achieved in a suitable form for administration by mouth this methenamine mandelate* is now one of our most useful drugs It is easily taken is non toxic and its action is independent of the bacterial population of the urine

The introduction of penicillin in its various forms during the last 10 years has given us yet another powerful weapon in dealing with those infections which are sulpha resistant. The disadvantage of the necessity for parenteral administration

*Mandelamine and Mandamine are registered trade names for this substance.

(except in tiny babies) has, however, limited the use of penicillin considerably, particularly in the treatment of chronic infections and in the prevention of recurrence *B proteus* still remained one of the greatest enemies of the urologist and the advent of streptomycin has to some extent removed this menace. It is yet too early to assess adequately the practical use of aureomycin and chloramphenicol in urological infections

Much has been written on the dangers of chemotherapy and antibiotic treatment, and it is important to realize the limitations of these treatments The early antibiotics, especially streptomycin, are bactericidal, whilst the sulphonamide group and certain antibiotics (aureomycin and chloramphenicol) are only bacteriostatic The natural defences of the body play a very large part in overcoming urinary infection We are coming to realize more and more that, especially in the urinary tract, organisms become resistant to a particular form of therapy There seems no doubt that a particular strain may become resistant itself, whilst on the other hand the urinary infection may persist by the replacement of a sensitive strain by an insensitive strain of exogenous origin There is now conclusive proof that not only do antibiotics cease to be effective against certain organisms but they actually stimulate the growth of other organisms It appears that penicillin may actually stimulate the growth of the tubercle bacillus and undoubtedly excites the rapid spread of pyocyaneus infections These 4 characteristics of organisms in relation to chemotherapeutic substances (suppression, inhibition, habituation and stimulation) (Garrod, 1951) must be constantly in the mind of anyone seeking to treat an infection of the urinary tract Constant vigilance, repeated chemical investigation of the urine and sensitivity tests on the organism present cannot be emphasized too strongly

Sulphonamide therapy

Sulphonamides have certain dangers which are, of course, greater where there has been damage to the renal parenchyma The effects of this group of drugs may be summarized briefly

Crystalluria and tube blockage—The solubility of drugs of the sulphonamide group varies considerably The amount of "sulpha" drug which is protein-bound also varies with the particular substance and this protein-bound fraction does not filter through the glomerulus The fraction of sulpha drug which, after filtering through the glomerulus, is re-absorbed from the tubules is also variable In the case of sulphadiazine, 65 per cent of the filtered sulphadiazine is re-absorbed, and in the case of sulphamerazine 85 per cent is re-absorbed Under normal circumstances only 1 of every 120 millilitres filtered by the glomerulus is actually passed as urine Consequently in the case of sulphadiazine 35 per cent of the sulphadiazine filtered is actually excreted in the urine but must be excreted in $\frac{1}{120}$ of the amount of fluid in which it was filtered by the glomerulus It is little wonder that crystallization occurs in the tubules with consequent tubule blockage

Nephro-toxic effect—It is uncertain whether the sulpha drug is actually cytotoxic or whether the effect seen in the kidney as a result of giving sulpha drugs is an allergic phenomenon Reference has already been made to pyelo-nephritis of allergic and toxic origin and the sulpha drugs are among the main precipitating

factors. Individual sensitivity of patients to sulpha drugs is of course well known and this allergy is shared by the renal substance Haematuria during treatment does not necessarily mean crystalluria but may be due to this allergic or toxic pyelo-nephritis. Necrosis of the epithelium in the distal convoluted tubule and collecting tubules has been found in many cases

Glomerular damage—Birchell and Alexander (1950) review the evidence which supports the occurrence of glomerular damage as a result of sulpha therapy and it appears that acute tubular necrosis may itself result from treatment with this group of drugs It has been shown that acute tubular necrosis often involves the glomerulus as well as the tubules

Allergy—A general allergic effect of a drug is, of course very important as febrile reactions can be very devastating and add to the dehydration and renal trouble already present. This allergy is specific to each drug and is related directly to the amount of the drug which has been given

Principles of chemotherapy

In severe infections of the urinary tract and in established chronic infection it is not enough to produce in the urine a concentration of the drug sufficient to deal with the organism and it is necessary to maintain an adequate plasma level so that the infection in the interstitial tissue in the kidney is also dealt with The effect of sulphonamide is directly dependent on the density of the bacterial population Since there is this direct relationship between the effect of a given amount of drug present and the actual bacterial population, it is logical (Garrod 1951) to attempt to reduce the bacterial population by the use of such other urinary antiseptics as are available before the exhibition of the sulphonamide group of drugs. By this means it may be possible to sterilize the urinary tract before resistance of the organism develops

After ingestion the fate of sulphonamides depends almost entirely on the characteristics of the particular member of the group although the body deals with all members along similar lines Solubility determines the rate and degree of absorption from the bowel After absorption a certain proportion of the drug becomes, by reaction with acetates, acetylated and this fraction of the drug is not bacteriostatic A further fraction becomes combined with protein and therefore fails to pass into the glomerular filtrate This protein combination is reversible so that the protein bound fraction remains as a store of drug in the bloodstream A small proportion of the sulphonamide is oxidized but the bulk is eliminated in the urine The rate of elimination by the kidney is again dependent upon the proportion which is not combined with protein The sulphonamide which is excreted is in two forms the acetylated and the non acetylated only the latter being bacteriostatic. Sulphacetamide itself is the only acetylated sulphonamide which is in fact bacteriostatic

As far as the urinary tract is concerned, therefore, the effectiveness of any member of the group will be highest if acetylation is slight and protein combination is low whereas a drug with high protein combination will last for a longer period and will also maintain a higher plasma level it will thus be more effective for infections involving the renal parenchyma

Most of these drugs are more soluble in alkaline than in acid medium, and for :s reason the urine is maintained on the alkaline side of neutral during therapy is, however, not necessary to do this if the amount of sulpha drug given is small, d when one is aiming at an effect in the urine and not at an effect in the tissues the kidney The exception to the general rule of an alkaline urine during atment is in the use of sulphathiazole against *Strep. faecalis*. The drug is then ›st effective when the urine is neutral or slightly acid Since the acetylated iction is most soluble, drugs with a high urinary clearance and low acetylation e most likely to produce crystalluria Sulphadimidine is 70 per cent acetylated t sulphathiazole has only 25 per cent acetylated in its urinary fraction

The renal excretion of the drug is dependent on the percentage of it which is otein-bound and not available for filtration Reference has already been made this fact If, therefore, one is treating pyelo-nephritis and requires a high plasma vel, it is better to use a drug with a high protein-bound fraction and high per-ntage of absorption from the tubules Sulphamerazine fulfils these conditions ı the other hand, with a simple recurrent pyelo-cystitis which responds to the esence of an antiseptic in the urine, it is very often sufficient to use a drug like lphacetamide which produces a very low plasma level and a high excretion rate small dose given at night and in the morning is excreted so adequately that a ry high urinary concentration is obtained

In determining the choice of drug it may be a help to consider the individual emical characteristics and biological reactions of some of the commoner members this group

Because of these various factors it has been found in practice that the use of a mbination of drugs is very often more effective and less dangerous than the rresponding amount of one single member of the group Each drug has its own rticular urinary solubility and this is independent of the presence of other drugs the same group It is therefore possible to maintain a fairly high urinary ncentration of total sulphonamide made up of several component fractions of e different members of the group It is also possible to maintain a high plasma vel and a high urinary concentration of sulphonamide by using a combination a drug with a high protein-bound fraction (sulphamerazine) and one of high lubility and lower tubular re-absorption such as sulphadiazine or sulphacetamide convenient preparation is

Sulphamerazine 0 1 gramme
Sulphacetamide 0 2 gramme
Sulphadiazine 0 2 gramme
in 0 5 gramme tablet form
or as a cream, 0 5 gramme in 5 millilitres
(Tresamide Sharpe and Dohme)

Some of the characteristics of the individual members of the sulpha group e given here as a guide to selection of the appropriate drug or combination of ugs

ılphanilamide—This drug is almost completely soluble at any physiologically ›ssible concentration, and it will be recalled that when these drugs were first troduced renal complications were not encountered until sulphapyridine came

into general use Sulphanilamide is however more toxic and less widely effective than many of its successors and it is almost non-effective against one of the most common infectors of the urinary tract—*Strep faecalis*

Sulphacetamide —This has the great advantage of being soluble in urine with a physiological acid *p*H and it is possible to achieve high urinary concentrations of the drug with a low plasma level A small dose, therefore, at night and in the morning will produce a very effective urinary concentration and will deal adequately with a urine infection by a sensitive organism not involving tissues deeper than the surface epithelium

Either of the last two drugs may be used with safety where the child is dehydrated and a positive fluid balance is difficult to achieve

Sulphathiazole —The main disadvantage of this particular substance is the greater tendency it has to produce allergic manifestations It is said to develop a concentra tion in renal tissue twice that found in the bloodstream or any other organ and, as has already been mentioned it is particularly effective against *Strep faecalis* and staphylococci especially with a *p*H below 7

Sulphadiazine —Less allergy and less nausea and vomiting are claimed for sulphadiazine, and only 14 per cent of the drug in the plasma is protein bound There is, therefore a fairly high glomerular filtration quantity, and 65 per cent is re absorbed from the tubules Owing to this fairly rapid clearance of the drug, crystalluria occurs fairly readily and both sulphathiazole and sulphadiazine are markedly nephrotoxic

Sulphamerazine (monomethyl sulphadiazine) —Sixty per cent of this drug in the plasma is protein bound and is therefore less effectively excreted Consequently a widely spaced dose gives a plasma concentration which is adequate for a longer period Eighty five per cent of the drug filtered by the glomerulus is absorbed from the tubules so that the excretion concentration in the urine is low It is therefore unlikely to be very effective in heavy infections It is also excreted by the renal tubules under certain circumstances The drug is almost entirely eliminated by the kidney and very little oxidized or destroyed in the body elsewhere The solubility of sulphamerazine is also low and in spite of the prolonged excretion time practitioners are in the habit of giving to children doses which are far too great and too frequent. We have personally seen too many cases of sulphamerazine urinary obstruction. The drug produces a bright yellow sludge in the bladder and these clumped crystals are responsible for acute retention by being impacted in the urethra (*see* page 2) Such an accident only occurs if the dosage has been excessive or too frequent One of us (T T H) first met this condition in 1945 on two occasions and Hawking and Lawrence (1950) make reference to it.

Sulphadimidine (syn dimethyl sulphadiazine, sulphamezathine) —This member of the group becomes more highly acetylated and is therefore less effective than the monomethyl derivative of diazine but is more soluble than either sulphamerazine or sulphadiazine if the urine is tending to be acid There is, therefore less likely to be crystalluria. In practice it seems to be an excellent drug for urinary infections, although it is said to be difficult to maintain adequate plasma levels even on 4-hourly dosage

A sulpha drug continues to be excreted in the urine up to 1 week a
administration has ceased Some members of the group are excreted fully in
first 48 hours and 75 per cent of sulphadimidine and 40 per cent of sulphamera
are excreted in 24 hours Sufficient may, however, appear in the urine (
succeeding days to prevent bacteriological culture of the specimen being of
value Direct microscopic examination and especially the clinical examinatio
the urine give a fairly reliable estimation of the success of treatment

Sulphamethazole (N U 445) —This is a sulphanilamide derivative (3–4 dime
5 sulphanilamide-isoxazole) It is very soluble in urine and is effective someti
against gram negative organisms which have become resistant to other drug
the group

The dose for a child of 6 years is 2 grammes initially and 1 gramme 8-ho
(0 2 gramme per kilogram body weight)

The drug was introduced in 1943, but has not been widely used (Fergusso
alii, 1948) It is now available (Gantrisin) for clinical use in Great Britain

The chemical test for sulphonamide in the urine

Occasionally, if the details of previous therapy are not known, a rapid test
urinary sulphonamide is useful This test (Bogen, 1943) depends upon the ef
of hydrochloric acid on cellulose (ordinary newspaper) which has been dippe
the urine to be tested A drop of hydrochloric acid (dilute 1 4) is put on a pa
of urine-soaked newspaper, an orange colour is given by sulpha drugs in
concentrations (25–50 milligrams per 100 millilitres) and a yellow by wea
concentrations (0 5–10 milligrams per 100 millilitres)

Antibiotic treatment

Since the introduction of penicillin, many workers have shown the rising numb
of organisms which are found to be resistant Barber (1947) showed that ove
period of 3 years the percentage of penicillin-resistant staphylococcal infecti
in one hospital rose from 14 to 59 per cent The clinician cannot, therefore, r
the continuance of penicillin therapy, if it is not immediately effective, with
insisting on sensitivity tests for the particular organism The great advantage
penicillin with infections which will respond is, of course, that there are alm
no untoward effects Allergic manifestations are extremely rare, though they
occur, and its prophylactic use during surgical management is of the utm
value Since the concentration of penicillin in the urine is extremely high, organis
which resist the effects of penicillin in concentrations found in the plasma m
succumb in the urinary tract, and *Strep faecalis* may respond to a urin
concentration of penicillin in the neighbourhood of 3 units per millilitre which
very easily achieved The other outstanding advantage of penicillin is that
action is independent of the urine acidity which, as we know, may swing from o
extreme to the other in a very short time

Streptomycin —The main use of streptomycin is in attacking the gram negati
organisms which are resistant to the sulphonamide drugs It is very rapi
excreted by the kidney, and with a distributed dose of 1 gramme a day in, f
instance, a child of 8 years, with a normal fluid output, it is relatively easy
obtain concentrations of over 1,000 units per millilitre, whilst many organisms a

susceptible to concentrations of less than 10 units per millilitre A simple urinary infection may disappear within a few hours of the first dose of streptomycin but against this is the very grave danger of acquired resistance on the part of the organism Streptomycin should therefore not be used in the primary stages of treatment for control of infection while surgery is pending or while there is still some degree of urinary stasis The weapon should be reserved for the final assault on the infection which may persist after all other methods of treatment have been completed Further it is necessary for the urine to be alkaline in order that streptomycin can be effective and in very small children it may not be easy to attain this

Allergic reactions to streptomycin have been described Other evidence of toxicity will not be found in the treatment of urinary conditions as the dosage needs to be so low In the long term treatment of tuberculosis of the urinary tract there is however a risk and deafness is perhaps the first evidence Aphthous stomatitis and hypertrichosis are also recognized complications of streptomycin therapy (Rouques, 1950 Fono 1950)

Aureomycin—Although an antibiotic, aureomycin is only bacteriostatic. Its use in the treatment of urinary infection should be reserved for those cases in which the organism is resistant to other agents but has been proved sensitive to aureomycin Dosage is rather empirical but may be given on the basis of 250 milligrams 8-hourly to a child of 5 years However owing to the nausea and diarrhoea which may occur the dose may have to be divided and spread over the day more evenly

Unfortunately the effects of aureomycin are not fully understood and severe and intractible *B coli* cystitis has occurred for no obvious reason in patients being treated by aureomycin for pulmonary and other infections

Chloramphenicol—The same restrictions in use apply to this particular antibiotic as to the use of aureomycin It is not bactericidal and sensitivity tests should be done before it is used The substance is used in similar dosage

Neomycin—Out of some 450 consecutive organism sensitivity tests, mostly in urinary tract infections, Duncan *et alii* (1951) found 62 strains completely resistant to penicillin streptomycin and Chloromycetin but sensitive to neomycin and they report considerable success in the treatment of urinary infections in a small series of cases

The dose which they recommend is 100 000 units 6-hourly for an infant continued for 3–9 days. The organisms which seemed to be most sensitive were the otherwise resistant strains of *Pseudomonas* and *A aerogenes*

The toxicity seems to be variable but albuminuria and tubule degeneration have been described and impairment of hearing by the effect of the drug on the acoustic division of the eighth nerve first may be sudden and severe. It would appear up till now that neomycin is probably too dangerous to use in its present form in the treatment of cases where the renal tissues may already be impaired.

The use of antibiotics and sulphonamides has become almost universal in the treatment of urinary infection but the hazards are not yet fully understood Penicillin and streptomycin are bactericidal but are selective in that they act on organisms which are dividing. If multiplication of organisms is prevented by the

use of a bacteriostatic agent such as a sulpha drug or even aureomycin, then there will be no dividing organisms on which the penicillin can take effect. Fortunately, in the urinary tract where there is a free flow of urine, a bacteriostatic agent is effective because the surviving inhibited organisms are washed out, but if drainage is not adequate cessation of the sulpha drug may lead to a revitalization of the infection. In spite of these theoretical considerations, in practice it is very often more effective to use drugs in combination than singly (Annotation *Antibiotic Synergism and Antagonism* (1950) *Brit med J*, 2, 1160)

Control of bowel infection

A severe bout of constipation sometimes precipitates a urinary tract infection, and in attempting to prevent recurrence of urine infection one should pay attention to the regulation of bowel function, particularly in those cases in which stagnation in the colon is severe Cauda equina lesions (spina bifida involvement) are invariably accompanied by gross constipation In certain cases, and in those children who have had the ureters implanted in the colon, we have gained the impression that urinary infection has been limited by the oral administration of low dosage sulphasuccidine or sulphathaladine The long-term effect of such bowel "sterilization" is liable to lead to interference with vitamin B synthesis, and is therefore of little practical value when more direct and safe methods of prophylaxis are available

In general practice, and in the treatment of a primary attack of pyelitis, it is clearly a matter of urgency to deal with the infection without waiting for bacteriological evidence, and the sulphonamides are undoubtedly the best weapon for this Success is often dramatic, the organism never being isolated. When, however, the infection is established and perhaps accompanied by some degree of obstruction, greater consideration must be given to the selection of treatment

SUMMARY

On account of all the chemical, bacteriological and biological factors which have been reviewed, it is therefore wise to plan a chemical attack on urinary infection so that the various agents may be used at the most appropriate time The promotion of a liberal diuresis and drainage, the early use of simple antiseptics, such as the mandelates, and attention to the nutrition and general condition of the patient will usually prepare the field for a much more effective attack by sulpha drugs or antibiotics During this phase, the sensitivity of the organism to the various agents may be determined and the appropriate sulphonamide drug selected We would reserve antibiotic treatment for "operative cover" and as a means of eliminating residual infection when the general plan of treatment has been carried out It is, however, impossible to lay down any rigid rules, for the organism may change frequently and consequently the treatment may have to change as well

When infection has been eliminated we believe that in cases of recurrent pyelo-cystitis, and certainly in children whose urinary tract is irrevocably damaged by dilatation, it is wise to insist on the continuous administration of Mandelamine with regular inspection of the urine Alternatively, sulphonamide therapy (for instance at age 7, 0 5 gramme night and morning) may be carried on without risk The urine is crystal clear normally when warm and acid, and if the parent notices

turbidity a full examination should be made Test papers* may be supplied to the parents for a regular rough check on the urine reaction

With careful observation and continuous preventive therapy many a child with a tendency to urinary infection may be steered through successive winters until past the tender years when we know these infections are so common and so devastating

PERINEPHRIC ABSCESS

Infection of the perirenal cellular tissue may be brought about by: (*a*) extension from the kidney from suppurative pyelo-nephritis, with or without calculus disease—this type therefore occurs with equal frequency in both sexes or (*b*) lymphatic or blood stream metastasis as in the well known staphylococcal abscess typically seen in male adults In childhood this variety is rare It follows



FIG 9—Operation specimen, left kidney and ureter removed from a girl aged 6 years.

At the age of 2 years this child had a urinary infection which persisted for 5 months, when she was admitted to The Hospital for Sick Children with left perinephric abscess. Urine heavily infected with B.C.C. Blood urea 148 mg. per 100 ml The abscess was opened and foul pus (B.C.C.) drained. Following this the child's general condition improved the blood urea fell to 20 mg per 100 ml., but the sinus in the loin was long in closing.

One month later intravenous pyelogram showed a silent left kidney right normal Cystoscopy demonstrated single U Os, the left gaping and discharging mucopus. On retrograde catheterisation the left ureter was demonstrated to be grossly dilated and the *renal pelvis small and contracted.* Later nephro-ureterectomy restored her to perfect health.

* Wide range indicator test papers are available (*British Drug Houses Ltd*) which are better as a guide than litmus paper

that in a perinephric abscess, associated urinary manifestations (such as pyuria, haematuria) may or may not be found With the aid of modern chemotherapy, suppuration can often be aborted and the condition resolved if recognized early The following illustrative cases may be quoted

Type (*a*) Fig 58 shows radiographically a perirenal abscess cavity occurring in an infant girl aged 6 months

In this case a large tender swelling developed in the right perinephric area, which resolved under treatment by penicillin Fig 9 illustrates a similar instance in which nephro-ureterectomy was ultimately necessary

Type (*b*) The following case occurred in the pre-penicillin era

A boy aged 10 years 11 months complained of severe pain in the left loin with fever and sickness On admission his temperature was 102° F There was fullness in the left loin, with acute local tenderness and muscular rigidity The urine was sterile W B C 16,900 per cubic millimetre

Four days later the swelling in the left loin had increased with a swinging temperature W B C 19,550 per cubic millimetre The loin was incised and a quantity of pus evacuated (*Staph aureus*) Recovery was uneventful

Pararenal abscess

Pararenal abscess may arise from an adjacent bony focus in the spine

B S Male Aged 3½ weeks

This infant was brought to hospital because a swelling had been noticed in the left loin The umbilicus was granulating and the urine contained a few pus cells and B C C On examination the left loin was obviously bulging and a lump, which felt firm and fixed, was found above the kidney, which could be distinctly palpated below and appeared reasonably mobile and of normal size The pyelogram showed the left pelvis displaced downwards Exploratory puncture under general anaesthesia drew a blank and a neoplasm was suspected The baby was immediately put on full doses of penicillin, however, and 2 weeks later the lump had practically disappeared One month later still no trace could be felt and the baby appeared perfectly well. Later radiological examination showed a small bony lesion in the body of L 1, and further pyelography showed the left pelvis in normal relation to the right Four years later this child turned up again with signs of a partial cord lesion, urinary incontinence. sacral anaesthesia, paralysis of glutei and calf muscles with bilateral calcaneovalgus Radiological examination of spine normal

PYONEPHROSIS

The renal pelvis may become urgently distended in acute infections, and relief by ureteral catheterization, direct aspiration or even nephrostomy may be necessary. The possibility that a true pyonephrosis may develop in a pelvis previously normal cannot be entirely ruled out, but there is no doubt that it almost invariably occurs in a pre-existing hydro-nephrosis

In the child gross pyonephrosis (empyema of the kidney) is most often seen in a subacute form, the initial illness, if noted at all, has usually been comparatively mild though some degree of local tenderness will usually have been noted Recovery is halted and the general condition gradually deteriorates until the child is obviously ill, sallow and anaemic, usually with a swinging temperature On examination a local tender lump is found in the renal area with a functionally silent kidney on pyelography Usually the wisest course in childhood is to drain

the abscess by nephrostomy postponing nephrectomy until the general condition has sufficiently recovered say 2–3 months later The removal of the kidney in these cases may be a formidable undertaking and it will often be found easiest to perform a subcapsular nephrectomy

Illustrative cases

Male aged 7 years

Had been unwell for 2 or 3 months getting worse No urinary symptoms had been noticed On examination the boy was obviously ill very pale, and a large mass could be felt in the left loin Daily temperature 98 -101 F Pyuria (profuse growth *B coli* and *Staphylococci*) B U 44 mg per 100 ml Hb 68 per cent Intravenous pyelogram showed right kidney normal left silent

After a small blood transfusion a large left pyonephrosis was incised, releasing a pint or more of stale greenish pus The general condition improved rapidly and the urine cleared One month later subcapsular nephrectomy was performed following which normal health was rapidly regained

On occasion immediate nephrectomy may be feasible as in the following nstance

Male, aged 5 years

Had been in increasingly poor health for 3 months with frequency and dysuria There appeared to have been no sort of acute onset but latterly some pain complained of in the right side, particularly on movement On examination the boy was pale listless and obviously ill There was an evening temperature of 100 F In the right loin was a large tender mass. Pyuria (*B proteus*) B U 35 mg. per 100 ml R B C 2,800 000 per cubic millimetre W B C 10 000 (75 per cent polymorph) Hb 48 per cent Intravenous pyelogram showed right kidney silent left normal

Following blood transfusion a large right pyonephrotic kidney was found to be sufficiently mobile to permit of an immediate nephrectomy The boy s post-operative condition caused no anxiety and 2 months later the urine was clear and st rile and the boy very fit

CHAPTER 6

EMBRYOLOGY

SINCE congenital anomalies provide much of the interest of children's urology, a study of the development is an essential preliminary From the embryology we can often get some idea of the range of possible variation, some rule for the irregular, every abnormality, however, must not be regarded as a simple "arrest of development" it may be that normal processes have been carried too far, or have been misdirected

In this section we will discuss in detail points of special interest to the paediatric surgeon, the broad outlines, which are well known and admirably described in many textbooks, are dealt with only in summary

TABLE 4

Germ-layer Origin of Urinary Organs

Organ	Origin
Secretory element of the kidney	Metanephros (mesoderm)
Collecting tubules Calyces Pelvis Ureter	Ureteric bud of the Wolffian duct (mesoderm)
Bladder and urethra	Urogenital sinus (cloaca) (endoderm)

Outline of development

The Wolffian ducts, mesodermal derivatives, first formed from the pro-nephric, later from the meso-nephric, tubules, grow caudally in the dorsal wall of the coelom until, reaching the hind end of the embryo, they turn sharply forwards and come into contact with the cloaca (4–5 millimetres) into which they soon open From the angle in the Wolffian duct the ureteric bud appears (5–6 millimetres) which, growing at first in a dorsal direction, is soon capped by a condensation of mesenchyme from the lower end of the nephrogenic fold—the primitive metanephros (*see* Fig 10). The ureteric bud elongates and bifurcates repeatedly while the metanephros is carried upwards from the sacral to the lumbar region from the ureteric bud are ultimately derived the collecting tubules, the calyces, the pelvis and definitive ureter, from the metanephrogenic cap, the nephrons.

The endodermal cloaca in a 6 millimetres embryo is the portion of hind gu lying caudal to the junction with the allantois, and cranial to the tapering tail gut. The cloacal membrane which forms the ventral wall of the endodermal cloaca is an area in which the ectoderm and endoderm are in direct contact As development proceeds the tail and the tail gut gradually degenerate and the sharp curvature of the hind end tends to unroll this brings the cloacal membrane into a relatively caudal position and at the same time an insinuation of mesoderm between its layers at the cranial end of the membrane separates the cloacal and umbilical regions to form the anterior abdominal wall Between 9 and 15 millimeters the cloaca is divided in the coronal plane by the down growth of the urorectal septum from the region of the junction of hind gut and allantois (Figs 11 and 12) the ventral division of the cloaca into which the Wolffian ducts open becomes the urogenital sinus, the dorsal division becomes the rectum The fusion of the urorectal septum

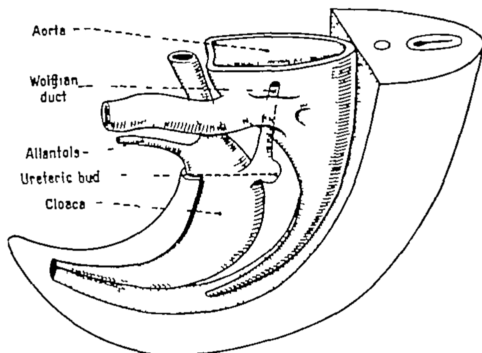


FIG 10—Reconstruction of a 5 millimetres embryo to show the relation of the Wolffian duct and ureteric bud to the cloaca and main vascular channels (after Keibel and Mall 1912)

with the cloacal membrane divides the latter into an anterior urethral membrane which ruptures (16–17 millimetres) soon after its formation and an anal membrane which persists considerably longer The intervening region thickens up to form the perineum

While these changes have been occurring in the cloaca the terminal segments of the Wolffian ducts have widened and are now gradually absorbed into the wall of the newly formed urogenital sinus The ureters then gain distinct orifices and are soon separated from the Wolffian ducts by the caudal movement of the latter the intervening region ultimately giving rise to the trigone (Fig 13) The cranial end of the urogenital sinus becomes widely dilated and forms the bladder (the allantois

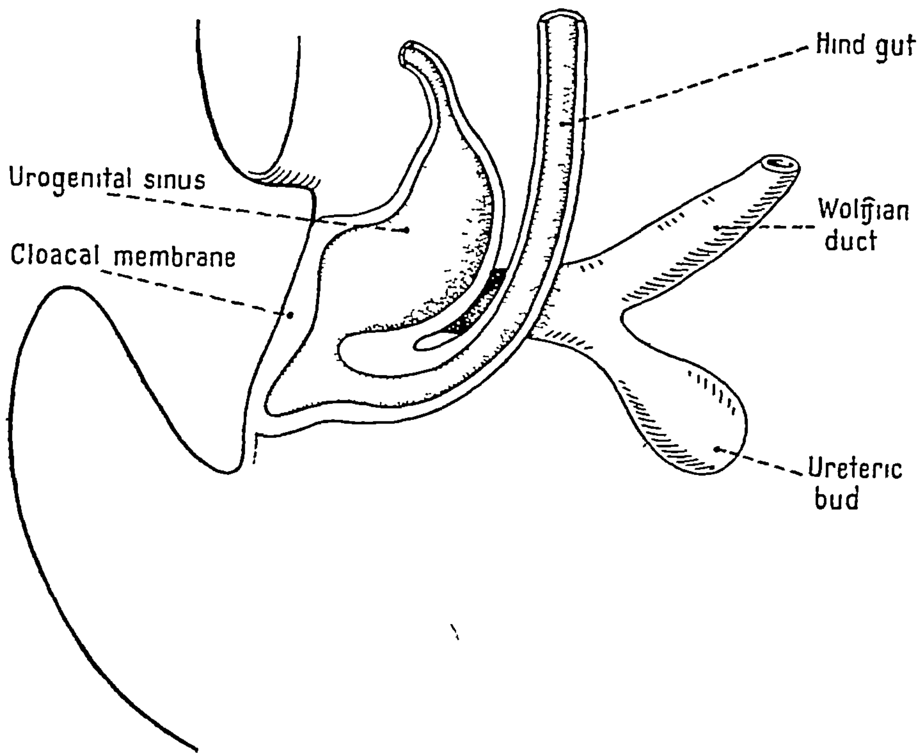


FIG 11 —The cloacal region at 9 millimetres showing the down-growth of the urorectal septum

Wolffian duct
Urogenital sinus
Cloacal membrane
Rectum
Ureter
FRANK PRICE

FIG 12 —The urogenital sinus at 11 millimetres showing completion of the urorectal septum and the broadening out of the terminal segment of the Wolffian duct

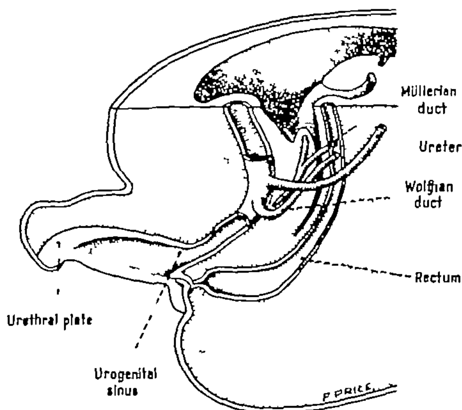


FIG 13—The urogenital sinus at 25 millimetres showing the loop on the Wolffian duct and the formation of the urethral plate adapted from Arey (1937)

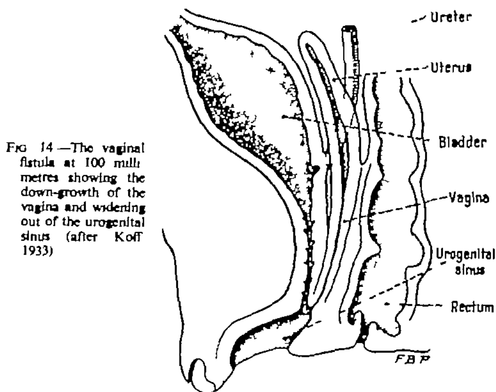


FIG 14—The vaginal fistula at 100 millimetres showing the down-growth of the vagina and widening out of the urogenital sinus (after Koff 1933)

degenerates completely), while the caudal end remains cylindrical and forms the greater part of, if not all, the urethra

The Wolffian ducts lose their urinary function in the male and come to subserve a genital one some of the mesonephric tubules join with the testicle and the ducts remain as the vasa deferentia In the female the Wolffian ducts normally degenerate and the genital passages are largely formed from the Mullerian ducts These appear in both sexes as invaginations of the coelomic epithelium covering the cranial end of the mesonephric folds (10 millimetres) and grow caudally Lying at first lateral, they cross anteriorly and come to lie medial to the Wolffian ducts in the condensation of mesenchyme behind the urogenital sinus known as the genital cord The caudal ends of the Mullerian ducts soon fuse and at 30 millimetres they have pushed up the posterior wall of the sinus in the region of the Wolffian orifices to form Muller's tubercle The upper bilateral portions of the ducts will differentiate as the Fallopian tubes, the fused portion as the uterus and upper vagina Muller's tubercle persists as the verumontanum in the male but in the female it is flattened out at 60 millimetres by two outgrowths of the sinus epithelium, the sino-vaginal bulbs The development of the female genitalia is complex but ultimately the sino-vaginal bulbs will form perhaps the lower fifth of the vagina, leaving the portion of the urogenital sinus which lay between Muller's tubercle and the bladder to form the definitive female urethra (Fig 14)

The external genitalia are largely derived from the mass of mesoderm interposed between the umbilicus and the cloacal membrane. A small hillock, the genital tubercle, is first evident in a 10 millimetres embryo, into this the caudal end of the urogenital sinus is prolonged and after rupture of the urethral membrane the urethral groove is to be seen on the caudal slope of the tubercle Closure of the lips of this groove will complete the male urethra and the genital tubercle becomes the penis. In the female no such closure occurs The lips of the groove remain as the labia minora, the urogenital sinus becomes shortened and widened until the vagina which arose in the neighbourhood of Muller's tubercle is open to the exterior through the vestibule and, as already mentioned, the female urethra ends at a point corresponding to the male verumontanum

The labio-scrotal swellings appear on either side of the genital tubercle at 17 millimetres As they develop they move caudally In the male they fuse in the mid-line at the time of the closure of the urethra and form the scrotum—the raphe remaining as evidence of their fusion, while in the female they grow together between the urogenital and anal areas as the labia majora and the posterior commissure.

The terminal segment of the Wolffian duct

A certain confusion of thought has been introduced into urological literature by the uncritical acceptance of the theory that the trigone of the bladder is formed from mesoderm of the Wolffian ducts and ureters Evidence against this theory was produced by Pohlman (1911), by Chwalla (1927a) and by Frazer (1935) A full account of the development of this region has recently been given by Gyllensten (1949), an account which is amply confirmed by our own observations

In the 10 millimetres embryo the terminal segment of the Wolffian duct opens as a funnel into the dorso-lateral horn of the urogenital division of the cloaca The

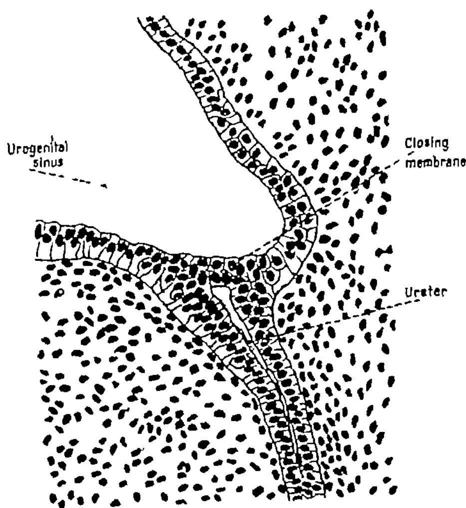


FIG 15—Cross section through the urogenital sinus of a 22 millimetres embryo showing the junction of the ureter and sinus occluded by the closing membrane.

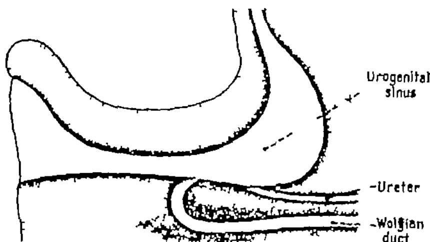


FIG 16—Diagrammatic parasagittal section through the urogenital sinus at about 24 millimetres, showing the terminal loop on the Wolffian duct and the absorption of the tissue lying between the lumen of the duct and the lumen of the sinus.

ureteric bud, previously a dorsal outgrowth, has now assumed a lateral position Between 10 and 14 millimetres the funnel gradually disappears, largely as a result of the outgrowth of the urogenital sinus epithelium into it, and although a part of the Wolffian duct may be taken up in the sinus wall, its epithelium is soon replaced In this way the ureter achieves a separate opening into the sinus, but no sooner has it accomplished this than its orifice is obliterated by a heaping up of epithelial cells A definite membrane is formed from these cells which continues to occlude the ureter until about 35 millimetres, that is to say, throughout the period during which the trigonal area is being formed (Fig 15) The ureteric epithelium at this time is also clearly distinguishable from the vesical, the cells being smaller and more deeply stained It is thus impossible to suppose that the ureter contributes to the formation of the trigone

In the meantime the Wolffian ducts with their widened ends have formed caudo-medial loops immediately proximal to their entry into the sinus Each loop is so placed that the final cranially directed limb is pressed against the wall of the sinus, and as the loop lengthens the intervening tissue between the lumen of the duct and the sinus atrophies and disappears (*see* Fig 16). This process continues so that the actual opening of the duct is moved caudally away from the ureteric orifice, leaving a groove in the dorsal wall of the sinus lined with Wolffian epithelium The groove is soon undermined by an ingrowth of the sinus epithelium and obliterated In this way the vertical extent of the trigone is formed, its breadth is accounted for by the dilatation of the upper part of the sinus to form the bladder, the ureters do not move apart from one another until this dilation is evident, and the whole trigonal area must be regarded as of endodermal origin

It seems likely that the beginning of the caudo-medial loop formation of the Wolffian duct is responsible for the apparent shift in the position of the ureteric bud from dorso-medial at 6 millimetres to lateral at 14 millimetres

The ascent of the kidney

It has been mentioned that when the ureteric bud is first found the metanephrogenic tissue which gathers around it is in a position which will correspond to the sacral region It is clear, therefore, that to reach the normal adult position a change must occur which may reasonably be described as ascent This process cannot, however, be regarded as an independent migration of the metanephros, nor is it likely that simple growth in length of the ureteric bud can push upwards its clumsy cap The "ascent" is complicated by, and perhaps partly due to, changes in shape of the organ during development

From the 6–10 millimetres stage the metanephros moves cranially along with the lower pole of the mesonephros, this movement is essentially the result of the straightening of the tail curvature which occurs at this stage and no active effort is required of the kidney The metanephros is then brought into contact with, and diverted by, the large umbilical arteries running forwards from the aorta to the abdominal wall (ultimately the common iliacs) so that the axis of the organ is shifted and the caudal pole brought up against the sacral vertebrae Growth of the kidney is now rapid and, since the caudal pole is fixed, takes place chiefly at the cranial end, with the increase in length the pelvis of the kidney soon reaches the level of the arteries and then passes them By 14 millimetres the cranial pole has



FIG 17.—Transverse section through an 11 5 millimetres embryo to show the convergence of the metanephros in the crutch of the umbilical arteries, the stage at which fusion might occur × 75

reached the lower border of the first lumbar vertebra, its definitive position After this elongated stage, the kidney again becomes rounded and the caudal pole is drawn up away from the sacrum At 22 millimetres the kidney lies alongside the first 3 lumbar vertebrae and subsequent changes in foetal life are only those due to variation in size of the adjacent liver and supra-renals During the first year of post-natal life the lower pole of the kidney frequently extends down to the iliac crest but the subsequent growth in length of the umbar region results in an apparent upward movement; the kidney itself is passive in this change (Gruenwald, 1943, Felix, 1912)

The metanephrogenic cap in the very early stages lies chiefly lateral to the tip of the ureteric bud but is soon shifted so that the ureter lies in front This relation persists until the kidney has reached the lumbar region, when a rotation occurs which brings the pelvis into its usual medial or anteromedial position During the ascent the metanephros makes use of a succession of mesonephric arteries in order to obtain its blood supply, the lower arteries degenerating after the upper ones have taken over

The pelvic ectopic kidney may be regarded as the result of a failure of this process of ascent, the ureter is short and springs from the anterior surface, and the blood supply is derived from the lower aorta or common iliacs The supra-renal glands, having an entirely separate origin from the genital folds, are normal in position, though they do not, of course, show the normal moulding

The fused kidney probably results from a defect at the 8–10 millimetres stage, when the two organs are pressed closely together between the two umbilical arteries (Fig 17) Should fusion be complete, further ascent will be impossible, and the "cake" or "disc" kidneys are pelvic in position If incomplete, the rapid growth of the upper poles may continue in the normal way, leaving the lower poles joined by an isthmus of renal tissue lying behind the ureters In these horse-shoe kidneys ascent proceeds up to the point where the isthmus is brought into contact with the inferior mesenteric artery, though on very rare occasions fusion of the upper poles, above the vessels, is observed In crossed ectopia—when both kidneys lie in the same loin, more or less fused, and the ureters enter the bladder in the normal position—the uncrossed kidney is often cranial to the other, and it has been postulated that fusion has occurred when one metanephros has ascended further than its fellow (Carleton, 1937), but in other cases the two kidneys lie side by side, and there are several anomalies on the side of the empty loin—hemivertebra, congenital dislocation of the hip, for instance (*see* Appendix III, case 4) These accompanying skeletal anomalies are also seen with the pelvic ectopic kidney, and though the connection between the two is not clear, it presumably indicates some early and fundamental defect of the mesoderm on one side of the body

The ureter

The early development of the ureter has already been discussed Up to 35 millimetres its lumen is narrow and its orifice occluded by a membrane. About this time secretion starts in the metanephros, the membrane disappears, and the lumen from then on constitutes about a third of its diameter It is surrounded by concentric layers of mesenchyme in which a few muscular fibres can just be made out The full musculature of the ureter, however, is slow to develop, forming from

below upwards and is not complete until about 150 millimetres (5 months)

After the fourth month the ureter shows a differentiation of form in 3 clearly defined segments The lower third lying caudal to the umbilical arteries has a circular cross section and a lumen held open by a firm muscle layer The middle third in the lower lumbar region appears as a flattened ribbon the lumen being almost obliterated by the apposition of anterior and posterior walls This segment is the most distensible if fluid is injected into the ureter but under normal circum stances it cannot properly be described as dilated. The upper and shortest segment forms an isthmus between the pelvis of the kidney and the lower lumbar spindle the muscle layer is here least well developed and the wall is frequently interrupted by a series of tight kinks or folds, which involve mucosa and muscularis but not adventitia The kinks have usually disappeared in the full term foetus and the ureter gradually assumes its uniform adult appearance A potential segmental pattern remains, however and may be shown up by pathological conditions The lower lumbar spindle is still the most easily dilated and when infection has rendered the muscle atonic ureteric dilatation is most obvious in this segment (*see* Lumbarureterectasis, Chapter 9) When the ureter is dilated from any cause, kinks tend to reappear in the upper lumbar isthmus and at the junction of the lower lumbar spindle with the pelvic ureter kinks which may be perpetuated by inflammatory adhesions Ostling (1942) has emphasized the manner in which the normal kinks of the foetal ureter may be responsible for the development of congenital hydro-nephrosis (*see* Chapter 10) In ureterocele and some types of megaureter dilatation first appears in the pelvic segment of the ureter a change by no means so reversible as the atonic dilatation of the lumbar spindle and indicative of a mild but long-standing obstruction

Congenital absence of the ureter is frequently accompanied by gross abnor malities of the Müllerian or Wolffian derivatives indeed it probably results from a primary failure of the Wolffian duct to reach the cloaca as has been observed by Boyden (1932) The trigone in such cases has a one-sided appearance but not infrequently the whole bladder is poorly formed on the side of the ureteric defect, and unilateral skeletal anomalies may also be present. Atresia of the ureter is more common than congenital absence There may be no more than a fibrous cord having a tenous connection with an aplastic kidney (*see* Fig. 20 page 79) Some of the cysts in the retroperitoneal region are attributed to dilatation of a segment of a ureter which has lost its lumen and perhaps its attachments at both ends others may represent mesonephric remnants

Duplications of the ureter result from the formation of an accessory bud either on the Wolffian duct or on the normal ureter A full description of the embryology of the duplications is given in Chapter 7

Retrocaval ureter is an anomaly resulting from an irregular formation of the inferior vena cava into which it does not seem necessary to enter here the ureter itself plays a purely passive role though it may of course become obstructed

The renal substance

Bifurcation of the tip of the ureteric bud takes place while it is still in the sacral region. From this time until the fifth month repeated bifurcation occurs with the formation of some twelve generations of collecting tubules Of these the third

and fourth generations dilate and are absorbed in the first and second to form the major and minor calyces, later generations becoming the definitive collecting tubules Differentiation of the metanephrogenic tissue is apparently induced by the proximity of the ureteric bud, in the absence of which the metanephros appears and even undergoes the first stage of ascent, but does not develop nephrons. Primitive glomeruli and uriniferous tubules are formed in connection with each generation of collecting tubules and, as the collecting tubules are gathered together in the renal pyramids, the grouping of the corresponding nephrons forms a surface lobulation on the foetal kidneys which disappears with continued differentiation in the cortical layer The uriniferous tubules related to the early generations of collecting tubules normally degenerate, but it is postulated that persistence of these structures may be responsible for the appearance of simple cysts or of polycystic disease in later life

The juxta-medullary glomeruli are naturally formed before the cortical with the repeated bifurcation of the collecting tubules and the glomerular tufts are covered during foetal life with a high columnar epithelium Both these factors are probably responsible for the small volume of urinary secretion by the foetal kidney, but there appears to be an unaccountable variation in this respect Many cases of complete congenital obstruction to the lower urinary passages have been described without any vesical distension On the other hand, gross dilatation of the bladder and ureter has occasionally been found in association with some minor obstruction, and no satisfactory explanation has yet been offered to account for this apparent difference in the volume of the urinary secretion The suggestion that the foetal kidneys may be provoked into activity by the partial failure of the maternal organs has never been adequately worked out

The bladder

The upper end of the urogenital sinus becomes dilated at the 40–50 millimetres stage and thereafter the demarcation between bladder and urethra becomes increasingly evident Dilatation also carries the apex of the bladder upwards to the umbilicus, so that the allantois plays no part in its development A condensation of mesenchyme has been gathering around the sinus from an early stage and by 50 millimetres muscle fibres have appeared in this Differentiation of musculature is usually stated to occur from the apex downwards, but circular fibres are evident behind the trigone as soon as anywhere, and longitudinal bundles running down between the ureters were observed in a 59 millimetres embryo The trigonal muscle does not appear until a later stage, but mesenchymal fibres from the lower end of the ureter can be seen running across the trigone at 94 millimetres.

Late in foetal life the apex of the bladder narrows down to form the urachal canal, which remains for a time attached to the umbilicus, but as the bladder descends into the pelvis with the growth of the child, the urachus loses its umbilical attachment though it may be held in position by the fibrous remnants of the umbilical arteries. The umbilical attachment may persist, however, and should there be an obstruction to the normal passages, urine may be discharged through the urachal fistula

Of the congenital abnormalities of the bladder only the apical diverticulum admits of an obvious embryological explanation other diverticula are normally

acquired as a result of urinary obstruction or at least are only congenital in so far as there is a pre-existing weakness in the musculature Symptomless diverticula have, however been observed in children without evidence of obstruction and Hyman (1923) has described cases of large sacculi causing ureteric dilatation in which the findings and follow up seemed to rule out any possible infra vesical lesion

Double bladder is so uncommon that a discussion of it is chiefly important to draw attention to conditions which may be mistaken for it It was early realized that large diverticula might be misleading, and Young (1926) suggests that in some cases the dilated lower end of a supernumerary ectopic ureter has caused confusion We have observed a case in which this difficulty arose (*see* Fig 44 page 102) The appearance of an hour glass bladder may be simulated by a large urachal diverticulum or by a dilated posterior urethra Chwalla (1927a) was of the opinion that a vertical partition in the bladder might be produced by the medial migration of the membrane normally occluding the ureteric orifice

Ectopia vesicae is the most interesting of the congenital anomalies of the bladder and is sufficiently frequent to constitute an important surgical problem The embryological theories are described in Chapter 14

The male urethra

The lower segment of the urogenital sinus, below the vesical dilatation is a roughly L shaped cavity the vertical limb the pars pelvina whose posterior wall has been completed by the junction of the urorectal septum with the cloacal membrane, and which receives the Wolffian ducts will form all the urethra down to the orifices of Cowper s glands in the bulb the horizontal limb the pars phallica consists of that forward prolongation of the cloaca which has from a very early stage extended up into the genital tubercle its caudal wall is formed by the urethral (formerly cloacal) membrane until 16–17 millimetres when this ruptures laying the sinus widely open to the amniotic cavity

Müller s tubercle a hillock in the posterior wall of the pars pelvina, is formed at about 30 millimetres by the fused tip of the Müllerian ducts pushing up in between the orifices of the Wolffian ducts Caudal to this the sinus shows a well marked crista urethralis posterior and a variable number of lateral submontanal folds. The prostatic tubules appear in the upper part after 60 millimetres by which time condensation of the surrounding mesenchyme already shows evidence of the formation of the external urinary sphincter The Wolffian ducts remain as the ejaculatory ducts Müller s tubercle as the verumontanum and the utriculus masculinus may perhaps represent the remnant of the Müllerian ducts though more probably it results chiefly from the sino-vaginal bulbs outgrowths of the sinus epithelium (*see* page 74)

At the time of the rupture of the urethral membrane the cavity of the pars phallica of the urogenital sinus does not extend as far as the tip of the genital tubercle but a solid lamella of cells continuous with the epithelium of the sinus is prolonged into the region now beginning to differentiate as the glans. In the shaft of the penis the open urogenital sinus shows as a groove which at the 40 millimetres stage is transformed into a tube by a fusion of its lateral margins (urethral folds) Within the glans, however the solid lamella of cells known as the urethral plate

remains unchanged and the plate can also be found dorsal to the newly formed urethra in the distal shaft (Fig. 18) The terminal urethra, the future fossa navicularis, is not completed until a much later stage together with the development of the prepuce.

The prepuce is first in evidence at about 60 millimetres as a solid ingrowth of cells from the surface of the genital tubercle in the region of the coronary sulcus This ingrowth, which is known as the glandal lamella, lies at first perpendicular

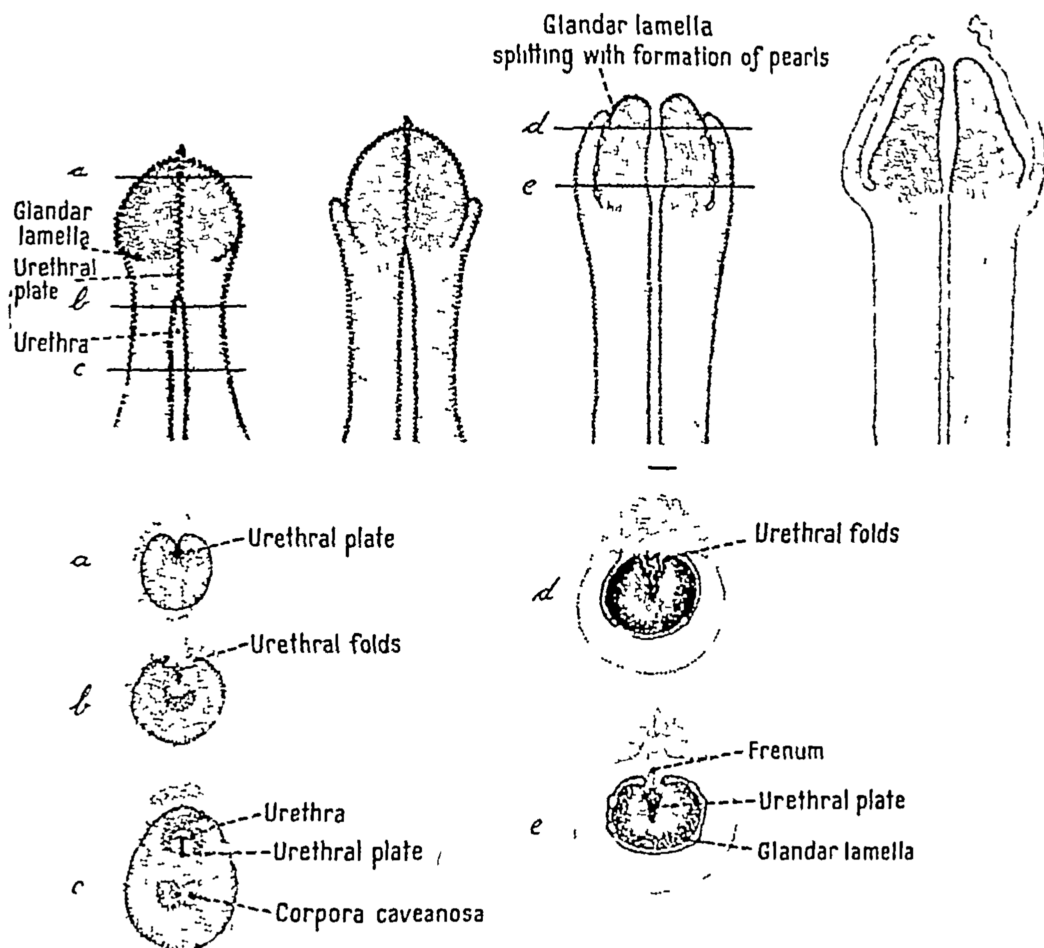


FIG 18—Diagram showing the development of the terminal urethra and prepuce The upper figures show coronal sections through the penis at 60 millimetres, at about 75 millimetres, at 100 millimetres and in post-natal life The lower figures represent transverse sections at the 60 and 100 millimetre stages

to the surface but later the growth of ectoderm covering the shaft of the penis pushes its point of attachment distally until the lamella itself comes to lie parallel to the surface and in the plane of the future preputial sac (Fig. 17) The glans may now be said to be covered by prepuce but the lamella does not split to allow separation of prepuce from glans until very much later indeed the process is still continuing during the first year of post-natal life Splitting of the lamella is preceded by the formation of epithelial pearls, clumps of degenerating cells, which are to be

seen when the prepuce of an infant is forcibly stripped back and which are some times mistaken for smegma The glandal lamella has from the first been deficient in the ventral mid line so that the prepuce here remains incomplete until the time (100 millimetres) when the fossa navicularis of the urethra is formed Fusion of the urethral folds in this region coincides with the fusion of the wings of the prepuce and the frenum remains to show the connection between the two processes

The solid urethral plate normally degenerates but partial separation and canalization accounts for many of the abnormalities found in this region from the common and scarcely abnormal lacunae of Morgagni and the valve of Guerin to the rare dorsal accessory urethral canals, which run back between the corpora cavernosa, and usually rejoin the normal urethra at the base of the penis

Hypospadias the commonest anomaly in this region may be regarded as a simple arrest of development, a failure of the closure of the urethral folds The orifice may be found in the coronary sulcus or at any point along the shaft of the penis but in cases where it is in the perineum the two labio-scrotal swellings also fail to unite, and on occasion there is a pseudovaginal pouch These cases of extreme hypospadias often present difficulties in the determination of sex, and are discussed in Chapter 16 Hypospadias is always accompanied by some degree of chordee (ventral curvature of the penis) partly attributable to the short urethra and failure of the corpus spongiosum The glans penis is tilted forwards because of the non union of the urethral folds in its ventral surface

The prepuce in hypospadias is commonly described as hooded in form it is however normal in shape except for the vertical defect which occurs through the failure of closure of the lateral urethral folds and the hooded appearance is simply the result of the retraction consequent upon this defect This fact is evident in neonatal cases before retraction has occurred, but the points at which the pre putial margin is interrupted by this failure of union are permanently marked upon the skin as two puckered dog-ears which can be found in all cases. The groove in the glans of hypospadiacs is often marked by 2–3 blind pits these result from the canalization of the urethral plate and represent the lacunae of Morgagni or para urethral ducts

Occasionally the hooded foreskin is found in association with a normally placed but stenosed, meatus in these cases since complete fusion of the urethral folds cannot have occurred the terminal urethra must presumably have been formed entirely by canalization of the urethral plate, and the stenosis is evidence of the incompetence of this method. Another variation is the split type of meatus under a normal foreskin the frenum is absent and growth of the prepuce must have resulted from a rolling forward of the ventral lip

Impermeability of the terminal urethra has been observed on a number of occasions (Ménégaux and Boidot, 1934) with and without vesical distension. In a few it has been associated with a patent fistula at the umbilicus.

Ventral cysts and diverticula of the penile urethra represent defects in the fusion of the urethral folds

The female urethra and lower genital passages

Up to 38 millimetres the development of the female urethra differs very little from that in the male. the urethral folds, however remain widely open, and from this

stage the sexes diverge At 60 millimetres the Mullerian tubercle is flattened out and destroyed by two outgrowths of the urogenital sinus, the sino-vaginal bulbs. The epithelium of the bulbs is stratified and the lumen soon obliterated by the mass of cells At the same time, the tip of the utero-vaginal canal (fused Mullerian ducts), which is pushed back by the bulbs, becomes stratified and solid The identity of the sinus and Mullerian derivatives is soon lost in the solid vaginal plate, which undergoes rapid proliferation At 150 millimetres the cranial end is demarcated by the formation of anterior and posterior fornices, while at the lower end the expanding vagina, now regaining its cavity, pushes in the posterior wall of the urogenital sinus and extends caudally (Fig 14) Meanwhile, the distal urethra has become shorter and wider, until the opening of the vagina is brought near to the surface in the shallow vestibule The hymen is formed from anterior paired elements derived from the urogenital sinus in the region of the original evagination of the sino-vaginal bulbs, and from a posterior median element, which represents the compressed posterior wall of the sinus caudal to the opening of the vagina

The proportion of the vagina derived from sinus and from Mullerian elements is hard to judge Koff (1933) attributes the lower fifth to the sino-vaginal bulbs It will be evident, however, that both sides of the hymen are ultimately derived from the sinus.

Complete absence of the female urethra is occasionally seen, with the bladder apparently widely open into the vagina, but in these cases it is hard to say what passage really represents the urogenital sinus and what the Mullerian ducts

Apparent valve formations in the female urethra are clearly not analogous to the valves in the male, and no theory can be put forward on embryological grounds to account for their appearance It is not uncommon to find a flap of mucosa growing forward from the posterior lip of the meatus, so that the urinary stream tends to be directed forwards, this does not always produce a stenosis, however, as was suggested by Boyd (1929), and we do not find it often a cause of urinary symptoms

It is tempting to find in the common adhesions of the labia minora a partial closure of the urethral folds, but the ease with which they are separated hardly supports this view

Abnormalities of the posterior urethra

Two abnormalities of the pars pelvina of the urogenital sinus are of the utmost importance to the paediatric surgeon—congenital valves in the posterior urethra, and atresia ani urethralis or the congenital recto-urethral fistula Viewed from the embryological standpoint, these two present important contrasts with the valves, although the effects on the upper tract are formidable, the primary anomaly represents a very slight variant from the normal, must occur late in development, and is seldom accompanied by any other congenital defect in the urinary tract On the other hand, atresia ani urethralis a gross displacement of the parts, results from an early deviation from the normal, and is frequently associated with other urinary abnormalities

Congenital valves—The great majority of cases of congenital valves (indeed all our clinical cases, and museum specimens to which we have had access) belong to

Young s Type 1 in which valves are formed distal to the verumontanum They are often bicuspid and the gap between them unless obviously the result of partial rupture, is anterior Their posterior margins are not infrequently continuous with ridges running down from the verumontanum though they may also appear to join together at the crista urethralis posterior far down the posterior urethra Consequent upon the ob truction the prostatic urethra is dilated the internal meatus is hypertrophied as part of the hypertrophy of the bladder muscle and appears as a sharp lip and a series of ridges more or less prominent, run upwards from the verumontanum to this lip

A study of the folds of the posterior urethra of the adult shows that they vary from a well defined median crista urethralis with no more than a suggestion of lateral ridges to a radiate arrangement of submontanal folds, arising from a little below the verumontanum and running laterally and forwards so that they almost encircle the urethra in the neighbourhood of the external sphincter In the urethra of the male foetus from 60 millimetres onwards these lateral submontanal folds can always be made out appearing in section to swing round from the crista urethralis to near the anterior mid line They vary considerably in their relative height, however and in one 99 millimetres embryo they were observed to be exceptionally well developed so that their upper ends surmounted the tall crista urethralis It is not difficult to conceive that an even greater development of the folds, with adhesion of their medial edges would result in the formation of typical valves A similar explanation was put forward by Lowsley (1912) after a study of the embryology of the prostate though he attributed the submontanal folds to the continuation of fibrous strands from the genital cord—strands which we have not been able to identify

Atresia ani urethralis—Although morphological theories have frequently been propounded to account for atresia ani urethralis and the arrangement of parts compared to the findings in the monotremes it seems more valuable for the surgeon to regard the defect as an atresia not altogether dissimilar to the atresia seen in other parts of the intestine Between 6 and 9 millimetres a normal atresia is occurring in the tail gut it is not impossible that this process may continue beyond its normal limits and affect that part of the gut destined to form the rectum Such an event would naturally interfere with the formation of the urorectal septum, and the rectum would be left opening into the urogenital sinus or attached to it by a cord Thus the common finding in these cases is a fistula a short distance below the verumontanum, but the contraction of the atretic gut pulls up and kinks the posterior urethra, and survivors usually show some degree of urinary obstruction The site of the verumontanum as will be seen from the preceding account of the development is determined by the site of the openings of the Wolffian ducts in the first place between which the Müllerian ducts intrude, and the caudal movement of the Wolffian openings (*see* page 66) may well be halted by the defect in the urorectal septum Thus the opening of the rectum usually lies caudal to the verumontanum. In the rare cases in which it lies cranial (atresia ani vesicalis) the gut is pushed out of the mid-line, and often accompanied by absent Wolffian derivatives on the affected side Rare cases have been reported (Spicer 1908) in which the rectum opened into

the mid-line of the bladder and the ejaculatory ducts lay on either side at the same level, and still on a level with the ureteric orifice.

In the female, it will be clear that the fistula immediately below Muller's tubercle will become a fistula in the fossa navicularis of the vestibule immediately below the hymen: such is, indeed, the ordinary finding. Recto-vaginal fistulae, however, are also seen in the region of the posterior fornix, and since the gut never has a normal junction with the Mullerian ducts, these openings must be regarded as a secondary development.

The dilatation of the urinary tract which accompanies agenesis of the abdominal muscles extends into the posterior urethra, but the nature of the obstruction there is not clear (Obrinsky, 1949, Housden, 1934, Daut, Emmett and Kennedy, 1947). The typical valve formations have not been described in this association. It seems possible, however, that the vesical distension is the primary disorder and the agenesis of the lower abdominal muscle in some way the result.

CHAPTER 7

CONGENITAL ABNORMALITIES OF THE UPPER URINARY TRACT

CLASSIFICATION

Abnormalities of the kidney
(1) Renal agenesis
(2) Renal hypoplasia
(3) Renal ectopia
(4) Renal fusion
(5) Renal cysts

Abnormalities of the ureter (duplications)
(1) Nomenclature and embryology
(2) The bifid ureter
(3) Complete pyelon duplex
(4) Complete pyelon duplex with ureterocele
(5) The ectopic ureter
(6) Triple ureter and diverticula

ABNORMALITIES OF THE KIDNEY

MOST of the anomalies found in the kidney are not in themselves productive of symptoms and when they are discovered during the routine investigation of minor urinary disorders a careful evaluation must be made before attributing to them an aetiological significance. Anomalous kidneys, however are unusually susceptible to pathological change and they have been described in association with every form of renal disease

Renal agenesis

In its bilateral form agenesis of the kidneys is of no concern to the surgeon. The child is either stillborn or dies soon after the placental circulation is cut off This anomaly is frequently associated with other congenital defects and with a somewhat characteristic facies (Potter 1946) (*see* Fig. 19)

Unilateral agenesis may be accompanied by absence of the ureter in which case the diagnosis can be reached cystoscopically It is certain that where there is no ureter there is never a differentiation of the metanephric tissue to form a kidney the adrenal having an entirely separate origin is unaffected. Absence of the ureter frequently results from an abnormality (perhaps absence) of the parent Wolffian duct, and an incomplete vas deferens or an abnormality of the Müllerian duct system such as a bicornuate uterus may be found in these cases Upon cystoscopy the corresponding ureteric orifice is absent and the whole trigone appears to be one-sided Sometimes the whole of one half of the bladder is underdeveloped, indicating a widespread interference with growth.

Where the ureter is present the lumen is commonly impervious at some point and it ends in a fibrous knot in the loin The lower part of the ureter may be a little dilated, or both ends may be sealed off leaving the middle segment to distend as a cyst Other abnormalities such as bifurcation or trifurcation may be found

The normal kidney contra lateral to an agenetic one is hypertrophied in compensation and its ureter is often somewhat dilated as compared with the normal

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Renal hypoplasia

Hypoplasia is more common than agenesis but it may be difficult to distinguish the congenital type from the secondary contraction of an inflamed kidney In its simplest form a unilateral congenital hypoplasia is represented by a small kidney with fewer pyramids than normal and fewer calyces, but having a smooth surface a normal histological appearance and a function proportional to its size



FIG 19—Facies of stillborn infant with renal agenesis (*By courtesy of Dr W E Hunt*)

Hypoplasia may also imply a fibro-cystic nubbin of tissue attached to the upper end of the ureter, in which no normal renal parenchyma can be found and which is without function (*see* Fig 20 (*a*) and (*b*)) The borderline between hypoplasia and agenesis is here hard to draw There appears to be no danger of hypertension developing in such an organ

A third group of cases is included under the term hypoplasia small nodular and cystic kidneys with some surviving normal tissue but a considerable degree of fibrosis (*see* Figs 21 (*a*) and (*b*) and 22 (*a*) and (*b*)) Some cases in this group undoubtedly result from an atrophic pyelo-nephritis in very early life, which has scarred the kidney and prevented further growth, in others evidence of present or past inflammation is not found, and it is presumed that a congenital inadequacy of the blood supply has caused the fibrosis The practical importance of the distinction between this type of congenital hypoplasia and atrophic pyelo-nephritis

is not very great. either when unilateral may be important in the development of hypertension (*see* Chapter 4) and when bilateral will result in chronic renal failure. Renal dwarfism of severe degree is frequently found with the bilateral form leading to death in later childhood or adolescence Ellis and Evans (1933) noted the frequent association of these kidneys with bilateral dilatation of the ureters and it has been suggested that the tremendous polyuria which accompanies

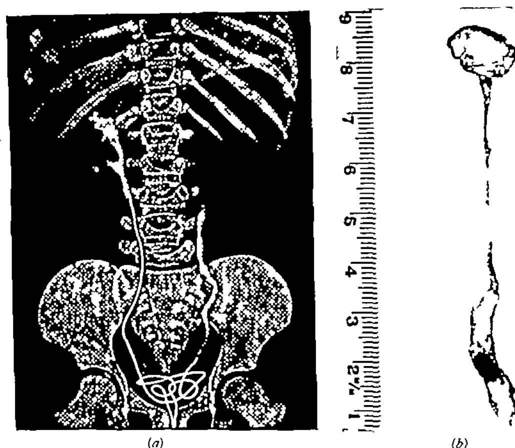


FIG 20 —Congenital renal hypoplasia. Female aged 6 years.
(*a*) Pyelographic tracing, (*b*) operation specimen.

Referred by Dr Sheldon for recurring urinary infection. I.V.P showed silent left kidney Retrograde pyelography demonstrated occlusion of left ureter Nephro-ureterectomy Satisfactory result.

the lesions is itself responsible for the dilatation As will be seen in Chapter 9 megaureter may be associated with a wide variety of lesions and the same infection which caused the atrophic pyelo-nephritis might also cause the ureteric dilatation. We are of the opinion that megaureter should not be ascribed to polyuria until all obstructive and infective possibilities have been eliminated.

The hypoplasia which sometimes accompanies moderate hydro-nephrosis is *almost always secondary to inflammation*

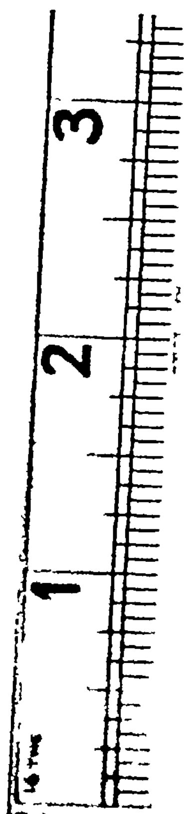
Both the unilateral agenesis and hypoplasia are important in contra indicating major procedures upon the opposite kidney Bell (1946) has estimated that of persons over 1 year of age 1 in every 207 has but a single kidney capable of sustaining life



(*a*)

FIG 21 —Renal hypoplasia
(*a*) Retrograde pyelogram,
(*b*) operation specimen

Boy aged 5 years 11 months, referred by Dr Pearson for recurrent haematuria Blood urea 46 mg per 100 ml BP 125/75 I V P showed R K silent, L K normal—rather large Note bulbous pelvis and rudimentary calyces right Three years later no further symptoms, BP 125/70





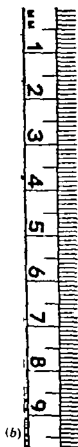
(*b*)



(*a*)

Fig 22.—Renal hypoplasia
(*a*) Retrograde pyelogram
(*b*) operation specimen

Girl aged 2 years. Recurrent urinary infection with occasional haematuria. Pyelography showed a poorly functioning malformed left kidney which was removed Infection took some time to clear but thereafter the child condition remained good BP normal throughout.





(*b*)

Unilateral ectopia and malrotation

The mechanism of the ascent of the kidney during embryonic life h described in Chapter 6, and it is not surprising that ectopic kidneys are cor found in the iliac fossa or in the pelvis All such organs are mis-shap commonly malrotated so that the pelvis projects from the anterior surf similar malrotation may be found in a normally placed kidney (*see* Fig 2 ureter may be short, but is sometimes long and tortuous, the arterial su



FIG 23 —Malrotation Pyelogram s malrotation of the left kidney T revealed by routine pyelography enuretic boy aged 5 years and bearing on the symptoms

derived from the lower aorta or iliac vessels, and several separate arteri usually to be found Ectopic kidneys are particularly liable to hydro-nep and to pyelo-nephritis They are not infrequently hypoplastic

In intravenous pyelograms the shadow cast by an ectopic kidney is apt overlooked as it is superimposed upon the sacrum (*see* Figs 24 and 25) enlarged, the mass in the iliac fossa is easily mistaken for an appendix absc a pelvic tumour

J W Male Aged 10 years (Referred by Dr Lightwood)

History 5 months previously had an attack of abdominal pain and vomitin which he was operated on at a local hospital for " acute appendicitis " The app was normal but a retroperitoneal mass was found and thought to tumour of the right kidney A further operation to remove this was undertake abandoned, the tumour being regarded as inoperable Following his discharge hospital the abdominal pains continued and the boy had occasional urinary i tinence

On examination a vague swelling was palpable in the right iliac fossa Intrav pyelogram showed a normal left kidney but no shadow on the right side Cystosc



Fig. 24

Fig 24 —Pelvic ectopia. Retrograde pyelogram showing left pelvic kidney in a girl aged 2 years, who suffered from vague but persistent abdominal pain and frequency of micturition Nephrectomy

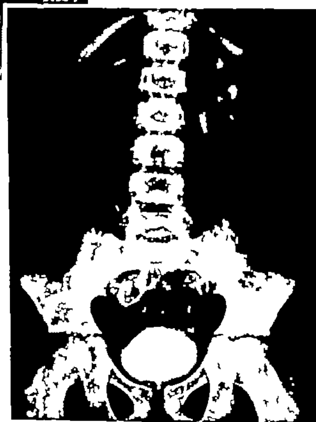


Fig. 25

Fig 25 —Pelvic ectopia. Excretion pyelogram showing right pelvic kidney in a boy aged 9 years who suffered from abdominal pain and enuresis. Nephrectomy Note the tortuous ureter and the complication of the bony background in radiological interpretation.

bladder normal Retrograde pyelography demonstrated a very large right pelvic hydronephrosis When distended with dye this stood out as a visible swelling in the right iliac fossa

Operation right paramedian laparotomy, many intraperitoneal adhesions Caecum mobilized and a very large hydro-nephrotic right kidney removed from the pelvis. The boy made a normal recovery and has had no further trouble

Where the contra-lateral kidney is normal there should be little hesitation in removing a diseased ectopic kidney Conservative surgery is difficult and unlikely to be successful Nephrectomy is best performed by the trans-peritoneal route, detaching the caecum or colon and drawing it medially to expose the organ.

Fused kidneys

The horseshoe type is much the most common form of fusion (*see* Figs 26 and 27), and even when not diseased may be associated with vague abdominal pains and



FIG 26—Horseshoe kidney Retrograde pyelogram showing left hydro-nephrosis in boy aged 5 years

The boy suffered from severe attacks of abdominal pain and vomiting associated with tenderness in the left loin Urine was sterile Intravenous pyelogram showed a horseshoe kidney with evidence of a hydro nephrosis of the left half At operation the cause of obstruction was found to be tethering adhesions of congenital type associated with an aberrant artery The obstruction was relieved by resection of the bands and division of the aberrant vessel plus a partial pelvectomy The boy had no further symptoms and has remained well for 8 years



FIG 27—Horseshoe kidney Intravenous pyelogram showing duplication of the left pelvis Note unusually developed upper segment

Boy aged 11 years of very small stature and with a minor degree of hypospadias His physique and mental condition were excellent and his build was familial The renal abnormality had no bearing

nausea The isthmus of the kidney lies below the origin of the inferior mesenteric artery and in a slim child it is frequently palpable where it crosses the vertical column The bodies of the kidney being low and anterior may also be felt even in the absence of pathology The pelvis of each half is rotated anteriorly and the lowest calyx lies medial to the ureter pointing towards the mid line. When both halves are functioning normally the pyelographic diagnosis is unlikely to be missed but hydro-nephrosis of one side is not uncommon and if it has entirely suppressed function a simple malrotation on the other side may be diagnosed Hydro-nephrosis is due to kinking by bands or vessels, or less commonly to compression of the ureter as it passes over the isthmus Conservative surgery should always



FIG 28—Crossed ectopia. Retrograde pyelogram from boy aged 5 years who suffered from chronic urinary infection and abdominal pain. Symptoms relieved by a meatotomy for a coincidental coronal hypospadias with meatal stenosis.

be considered in the case of hydro-nephrosis of a horseshoe kidney since the other side can never be said to be normal In two of our cases we have found considerable improvement in function following the release of the ureter from adhesions and from the compression of a vessel running to the lower pole When the isthmus is suspected as the cause of the obstruction it may be divided without difficulty and a nephropexy should then be performed on the side of the hydro-nephrosis Where advanced hydro-nephrosis, calculus disease, tuberculosis or neoplasm indicates removal of one half of the organ hemi nephrectomy presents no particular difficulty

The fused pelvic kidney is less common and is seldom amenable to surgical treatment. It is occasionally palpable as a mid line swelling, which when associated

with abdominal pain may give rise to difficulty in diagnosis. Pyelography will, of course, reveal the nature of the condition.

In crossed ectopia, where one kidney has migrated to the opposite loin, the two organs are almost always fused and the ectopic kidney lies below the normally placed one. *The ureters enter the bladder in the usual positions* (*see* Fig. 28). As with the fused pelvic kidney, surgical treatment is only of value where there are obstructing calculi which may be removed by pyelotomy (*see* Appendix III), case 4.

Cysts of the kidney

A few cysts are commonly found in hypoplastic kidneys and where lower urinary obstruction has started early in life cortical cysts, as well as hydro-nephrosis, are likely to be present in the kidney. Simple cysts of the type formerly known as "solitary" and occurring in an otherwise normal kidney are exceptionally rare in children and need scarcely be considered in the differential diagnosis of a renal tumour.



FIG. 29.—Congenital polycystic kidney. Excretion pyelogram from a boy aged 4 years who presented with a lump in the right loin. A renal embryoma was suspected.

There was in this case a familial history of congenital polycystic kidney and at an exploratory operation the nature of the lump was confirmed as such. Nothing further was done. (By courtesy of Garrod, Batten and Thursfield, and Edward Arnold and Son.)

Polycystic disease is seen in infancy and very rarely in older children. The infants are stillborn or die of renal failure soon after birth. The size of the kidneys in the infantile cases varies very considerably from small organs which might be considered hypoplastic, through normally sized kidneys which are honeycombed by small cysts hardly distorting the normal outline, to large irregular cystic masses similar to the kidneys of the adult disease. Other abnormalities of the urinary tract, or of its blood supply, are common in the infantile group.

In later childhood the adult type of polycystic disease is occasionally encountered. One kidney not infrequently enlarges before the other and if this enlargement is accompanied by haematuria differential diagnosis from renal tumour can only be made by a careful study of the retrograde pyelogram (*see* Fig. 29) The progress of the disease is seldom sufficiently rapid to cause renal failure during childhood years, but a case of hypertension has been recorded Complicating pyelo-nephritis and even stone are described



FIG 30—Unilateral multicystic disease. *Operation specimen from* male infant aged 5 weeks. Presented with mobile lump in right iliac fossa. Removed by laparotomy the left kidney having been palpated and found normal.

There is increasing evidence for separating as two distinct diseases the adult and the infantile form Lambert (1947) in a careful study of serial sections, has shown that in the adult disease although glomerular cysts occur the majority of cysts are formed in the course of patent and functional tubules whereas in the infantile form tubules attached to the cysts end blindly Fergusson (1949) points out that in the adult form a clear familial history is almost always forthcoming, but although the siblings of the infant affected may have died of the disease it is not found in the parents or more distant relatives Other congenital abnormalities of the urinary tract are common in the infant but very exceptional in the adult.

With regard to the aetiology of the cysts the classical theory of a failure of union between the nephrons and the collecting tubules is not altogether satisfactory and Kampmeier s hypothesis that the cysts represent the earlier generations of collecting tubules, which normally degenerate, has more in its favour, particularly for the adult type. In the infants, Lambert s work has shown that the failure


(*a*)



FIG 31—Unilateral multicystic disease Boy aged 5 years (*a*) Retrograde left ureterogram note displacement of the ureter across the mid-line (*b*) operation specimen showing hydro-nephrosis associated with parenchymal cysts one very large, without obvious connection with the pelvis

The patient suffered from attacks of vomiting and pain A lump of considerable size was palpable in the left loin Urine was sterile Blood urea 35 mg per 100 ml BP 110/80 Intravenous pyelogram showed a normal right kidney left side silent

of union has in fact occurred, but this author is of the opinion that the failure is a result rather than a cause of the cyst formation

A unilateral polycystic disease is occasionally encountered in children but probably has no connection with the bilateral form and is perhaps better known as multicystic disease. Ravitch and Sanford (1949) describe 4 cases in young infants with absent or impervious ureters and complete destruction of the kidney by large cysts We have encountered one such case and another in which a moderate hydro-nephrosis was accompanied by large cortical cysts having no obvious connection with the pelvis (*see* Figs 30 and 31 (*a*) and (*b*) In the latter the ureter was completely obstructed at the pelvi ureteric junction though in view of the hydro-nephrosis, this was presumably a secondary effect These cases with cysts were distinguished from simple hydro-nephrosis by the extreme displacement of the ureter across the mid line of the body which is not seen in uncomplicated hydro-nephrosis in children

ABNORMALITIES OF THE URETER

Duplication

A considerable number of children investigated for serious urinary infection or other manifestation are found to have duplications of the upper urinary tract. The anomalies may be on one or both sides and exhibit every variety of type and degree and the complications can be many and various The subject is therefore one of great practical importance in the urology of childhood

Nomenclature

In describing the duplications it is convenient to use a term which will embrace all the derivatives of the ureteric bud (that is ureter pelvis calyces and collecting tubules) and for this purpose we employ the word pyelon * *Pyelon simplex* will then describe the normal condition of the upper tract, while *pyelon duplex* will cover all forms of duplication

Embryology

A reference to the embryology is necessary to show the connecting link between the various types of duplex and to make clear the anatomical relations Fig. 32 shows very diagrammatically the normal development of the pyelon simplex which has already been described in Chapter 6 The ureteric bud arises from the elbow of the Wolffian duct where this turns forward to join the cloaca The segment of the duct caudal to the elbow disappears by widening out and being incorporated in the wall of the urogenital sinus (cloaca) After this process is complete and the ureter has attained a separate opening into the urogenital sinus the extremity of the Wolffian duct forms a caudally and medially directed loop which by absorption of its terminal limb brings the orifice of the duct into a position caudal and medial to the ureteric orifice that is in o the definitive position of the ejaculatory ducts (*see* page 66)

Pyelon duplex —This is the result of the formation of an accessory ureteric bud either on the side of the normal ureter or on the Wolffian duct itself and the exact

* η πυελο —A receiving vessel

Fig. 32.—Normal development of ureter and Wolffian duct. The ureteric bud arises from the elbow of the Wolffian duct. The terminal segment of the duct widens out and is absorbed into the urogenital sinus. The remaining portion of the duct then forms a caudo-medial loop, which eventually brings its orifice into a position caudal to the ureter (*see* pages 64–66)

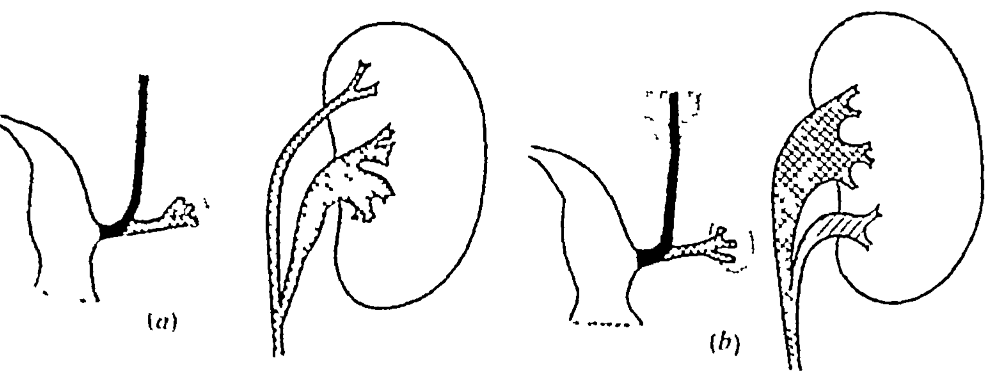
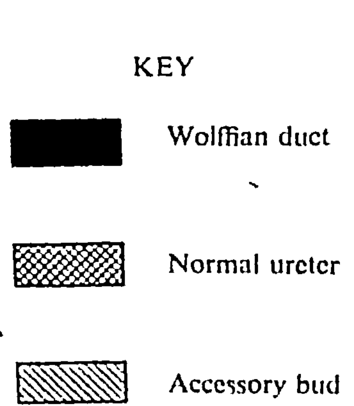


Fig. 33.—Incomplete pyelon duplex. (*a*) Shows an accessory bud formed on the cranial side of the normal ureter, and the common type of bifid ureter; (*b*) shows a caudal accessory bud—the rare type



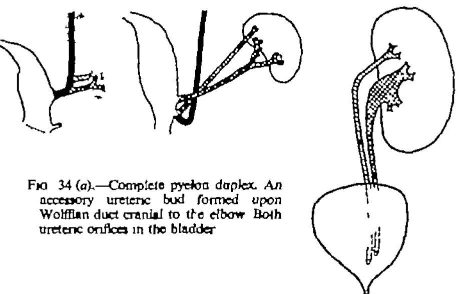


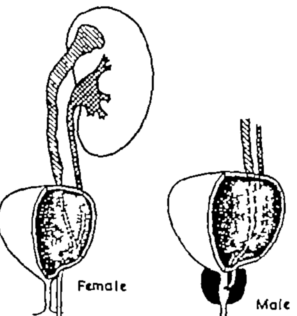
FIG 34 (a).—Complete pyelon duplex. An accessory ureteric bud formed upon Wolffian duct cranial to the elbow Both ureteric orifices in the bladder



FIG 33 —Abortive accessory bud. An accessory ureteric bud formed upon the Wolffian duct caudal to the elbow In this position it will probably degenerate



FIG 34 (b) —Complete pyelon duplex with ectopia. An accessory ureteric bud formed upon the Wolffian duct cranial to and at some distance from, the elbow The orifice of the deutero-pyelon is brought by the caudo-medial loop of the Wolffian duct to a point outside the bladder



site of origin of this accessory bud is of great importance in determining the final anatomical relations There can be little doubt, however, that the same embryonic "inductor" is responsible for all types, since the bifid ureter and complete duplex are frequently found on different sides of the same subject, and there is a continuous series bridging the gap between the two extremes—double pelvis and complete duplication with ectopia

It follows from the double nature of the bud that all its derivatives will be separate accordingly double ureter is accompanied by double pelvis, and the calyces of the two systems scarcely ever communicate The mass of metanephric tissue, on the other hand, may remain single although the nephrons are being differentiated in relation to two systems of collecting tubules Thus the double kidney, though commonly longer than the normal, shows on its surface very little evidence of its duplicity. There may be a sulcus, however, which indicates the junction of the two segments, or very rarely there is complete separation and the smaller mass is referred to as a supernumerary kidney. The blood supply to the two segments is very variable (*see* page 68)

Fig 33 (*a*) shows the formation of the bifid ureter which results from an accessory bud arising from the cranial side of the normal one This accessory bud comes into contact with the upper pole of the metanephros, and in the definitive kidney the upper calyces drain into the small upper pelvis the lower and more normal pelvis is formed from the original bud Fig 33 (*b*) shows the rare condition in which the accessory bud arises on the caudal aspect of the main formation The site of the bifurcation of the ureter will clearly depend upon the site of origin of the accessory bud and there may be simply a bifid pelvis, or the fork may lie at any lower point along the length of the ureter and even actually within the bladder wall In the latter case the two ureters may lie parallel to one another, but more often crossing in the frontal plane can be seen Very rarely one ureter makes a complete spiral turn round the other—an arrangement which cannot be reconciled with the usual formation of an accessory bud, and which results perhaps from the formation of a spiral septum

In the complete pyelon duplex it is convenient to refer to the derivatives of the normal bud as the protopyelon and to those of the accessory bud as the deuteropyelon

Fig. 34 (*a*) shows the common type of complete duplex in which the accessory bud has formed on the Wolffian duct, immediately cranial to the elbow So placed, the bud makes contact with the upper pole of the metanephros and results in a kidney indistinguishable from that found in association with the common type of bifid ureter but it will be seen from Fig. 34 that the caudo-medial loop of the Wolffian duct brings the orifice of the deuteropyelon into a position caudal and medial to that of the protopyelon, that is to say to a position on the trigone nearer the internal meatus. Again, the ureters may lie parallel, but frontal crossing in two places is the rule The two are bound together by a loose sheath throughout their length, but in the lower half-inch, besides being enclosed in a common "sheath of Waldeyer", they are much more closely adherent and separated by sharp dissection

Fig 34 (*b*) shows what will happen if this type of [illegible] the Wolffian duct at some little distance cranial to the

a bud will not make a satisfactory junction with the metanephros, and the resulting calyces are poorly formed, while the protopyelon is left free to develop upon almost normal lines The loop on the Wolffian duct will carry this bud further caudally so that the deuteropyelon may open at the bladder neck in the posterior urethra, or may fail to reach the urogenital sinus altogether Thus in the male, this ectopic ureter may end in the ejaculatory ducts the seminal vesicle or the vas deferens (derivatives of the Wolffian duct) whereas in the female the degeneration of the parent duct may leave the ureter to open into the vestibule or to form a fistulous connection with the vagina, uterus or even the rectum

The musculature of these abnormally derived ureters is often defective and in clinical cases the ectopic ureter is always dilated at least in part On occasion the ectopic ureter is the only ureter on that side and presumably no bud has arisen in the normal position at the elbow (ectopic pyelon simplex)

The lower end of the ectopic ureter may lie very close to the trigone and to the lumen of the urethra. in one of our cases no proper muscle layer intervened between the two In other instances the deuteropyelon ends blindly or through a narrow orifice and the dilated lower end bulges into the bladder as a ureterocele or ureterovesical protrusion In the cases in which it accompanies complete duplex the ureterocele is always found upon the deuteropyelon doubtless because of its longer intramural course and anomalous termination The ectopic ureter and the ureterocele with duplication are more common in the female than in the male

Fig. 35 shows the formation of an accessory bud on the Wolffian duct caudal to the elbow (this has been described in a 10 millimetres embryo by Chwalla (1927b)) In this position it is unlikely to reach the metanephros and accordingly we have been unable to find a description of a complete duplex in which the cranial pole is the main formation It may be argued that an accessory bud in this position is responsible for the formation of the congenital vesical diverticulum, behind and lateral to the normal ureteric orifice Since however the section of the Wolffian duct caudal to the elbow disappears as a result of widening out and being taken up into the urogenital sinus and takes no part in the loop formation, we believe that an accessory bud in this position would be lost altogether and could not attain a position outside the trigonal area

Incomplete pyelon duplex (the bifid ureter)

Diagnosis

Apart from those discovered only at operation all cases of bifid ureter are diagnosed by pyelography The lower pelvis is the larger and more normal in shape, though it can usually be seen when the upper half is not outlined that the upper calyces are missing, and that the outline of the pelvis slopes downwards immediately on leaving the renal substance and shows none of the normal projection towards the mid line The upper pelvis is small and consists of little more than two minor calyces the ureter is directed at first downwards and medially then turns downwards at a rather well marked angle (*see* Fig. 33 (*a*) (diagram) and Fig 36) Occasional variants on this characteristic pattern are seen the two pelves may be equal in size and similar in shape or very rarely the upper one may predominate The bifurcation of the ureters may occur at any point between the kidney and the bladder and is often hard to

site of origin of this accessory bud is of great importance in determining the final anatomical relations There can be little doubt, however, that the same embryonic "inductor" is responsible for all types, since the bifid ureter and complete duplex are frequently found on different sides of the same subject, and there is a continuous series bridging the gap between the two extremes—double pelvis and complete duplication with ectopia

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Incomplete pyelon duplex (the bifid ureter)

Diagnosis

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make out in the pyelogram, lateral views can be of great assistance in determining the site Where retrogade pyelography is being performed, both ureters may be filled if the dye is run in while the catheter is just within the orifice, with the catheter pushed up to the kidney, clearly only one pelvis will be shown



FIG 36—Incomplete pyelon duplex Bifid ureter Pyelogram showing typical appearance of an incomplete pyelon duplex on the left side in a girl seen for recurrent infection, controlled by conservative means

In our experience this variety of duplication is specially prone to serious infection The point of bifurcation may constitute a point of obstruction to one or both ureters, the upper being a little more often involved The lower, more normal, pelvis is less frequently affected in this way (*see* Figs 37 and 38)

Complete pyelon duplex

The deuteropyelon having a separate opening on the trigone (Fig 34 (*a*))

Diagnosis

The pyelograms in these cases are similar to those seen in the incomplete pyelon duplex with a small upper and large lower pelvis It may be possible to trace the outline of both ureters throughout their length, but a low bifurcation can seldom be excluded by radiological examination alone Upon cystoscopy, two ureteric orifices are seen on the same side usually one lies at the corner of the trigone (protopyelon, lower pelvis) while the other (deuteropyelon, upper pelvis) is found at some point nearer the internal meatus on the lateral border of the trigone Occasionally the two orifices may be side by side, the medial always representing the deuteropyelon, and cases have been reported in which the latter was actually



Fig. 37

FIG 37 —Incomplete pyelon duplex. Bifid ureter with infection and dilatation of upper segment in girl aged 8 years. Excretion pyelogram. note absence of secretion upper pelvis left and unusually small lower segment.

This girl complained of bdominal pains, frequency and haematuria. On examination the left kidney was palpable and the urine purulent. Cystoscopy—both UOs normal. A retrograde pyelogram demonstrated a dilated upper seg ment on the left side but did not fill the lower The point of junction of the ureters was not clear but was evidently near the bladder Hemi-nephro-ureterectomy was performed. Result—cure.



Fig. 38

38 —Incomplete pyelon
uplex (right) with infec
on and dilatation of
wer segment. Retro-
ade pyelogram from
fant boy aged 1 year 10
onths. Nephrectomy
rine cleared at once.

a little further from the meatus than the protopyelon A search for a second orifice is an important part of any cystoscopic examination in the child. unless the observer is very much on the alert, the anomaly is liable to be overlooked even in the uninflamed bladder In the presence of cystitis, the identification of the second orifice may be impossible. While the child with complete pyelon duplex is liable to troublesome infection, this is more easily controlled than in the incomplete type, and the prognosis is therefore more favourable Serious complications are, however, not infrequent.

Pathology

In this type of duplication, it is not uncommon to find considerable dilatation of the protopyelon, that is an infected megaureter (Megaureter (Group 6) *see* Chapter 9). The deuteropyelon is seldom involved except by the pressure of its fellow The cause of the dilatation and its selectivity is not clear there is no obvious obstructive factor The symptoms of this type of megaureter do not differ from others in which no obstruction can be found They are commonly those of pyuria with perhaps an aching pain in the flank The treatment of infections may be conservative, but once serious dilatation has appeared surgery will probably be required Hemi-nephrectomy may be practicable but the remaining upper pole may be so small as to be scarcely worth preserving

Complete pyelon duplex with ureterocele

As already explained, in complete duplications (Fig 34 (*b*)) a ureterocele may be found upon the lower orifice, that is the deuteropyelon Because of its origin, it



FIG 39 —Specimen in the Museum of The Hospital for Sick Children Complete pyelon duplex right with ureterocele

is nearer the internal meatus than a ureterocele on a pyelon simplex, and therefore more apt to obstruct the outlet of the bladder (Fig. 39) In some cases the cystic lower end of the deuteropyelon appears to be blind, at least no orifice can be found and such cases are closely related to those with an ectopic ureter and dilated lower extremity (intravesical protrusion Figs 40 (*a*) (*b*) and (*c*) and Appendix III cases 1 and 2)

Diagnosis

Dilatation of the upper p lvis may be seen in the intravenous pyelogram but it is more usual to find this segment silent owing to its defective function and only the lower pelvis l outlined Upon cystoscopy a cystic swelling balloons into the bladder from one side of the trigone and may appear to extend down to the internal meatus The stenosed orifice of the deuteropyelon may be seen on the near surface of this swelling the orifice of the protopyelon lies on the upper surface often dragged up by the distension of the cyst The contra lateral orifice is dis placed towards the side of the lesion and may appear to be held open If the ureterocele has been responsible for vesical neck obstruction the bladder may be trabeculated The swelling may be felt bimanually under anaesthesia with the bladder empty It is seldom possible to catheterize the orifice of the deuteropyelon but the ureterocele may be punctured with a diathermy electrode and a catheter inserted into the hole so formed Retrograde pictures can be obtained in this way (*see* Fig. 40 (*b*))

The ectopic ureter

Alldred and Higgins (1951) have recorded 4 cases 1 of them bilateral of this anomaly which passed through the urological service at Great Ormond Street in a 3 year period. The clinical features are easily missed during early childhood and the characteristic incontinence is treated as enuresis

The ectopic ureter is usually derived from the upper pelvis of a double kidney It may however be the only ureter on that side, in which case the kidney itself is often abnormal and the corresponding half of the bladder may be missing. Debenham (1950) has reported such a case (*see* Fig. 41)

In the male the ureter may end in the posterior urethra, where it can be seen urethroscopically or in the ejaculatory duct or seminal vesicle We record one case of ectopic ureter in the male which was characterized by pyuria and some obstruc tion to the bladder neck resulting from the dilated lower end incontinence was present but it was not attributable to the same cause as in girls (*see* Appendix III case 2)

In the female the extravesical opening may be in the urethra at the external meatus in the vestibule (*see* Fig. 44) the lateral vaginal wall, or even in the uterus In all these cases it may be the cause of a characteristic incontinence—the continual dribbling of urine despite micturition at normal intervals In our cases the incon- tinence has usually been exclusively diurnal or only when the child sits up in bed, that is the vertical incontinence to which reference was made in Chapter 1 In the recumbent position the ureter is sufficiently flaccid to form a *reservoir* of urine which on y escapes under the influence of gravity

The upper part of the ectopic ureter is always dilated and the pyelogram seldom shows any recognizable pelvi ureteric junction In some cases the dilatation

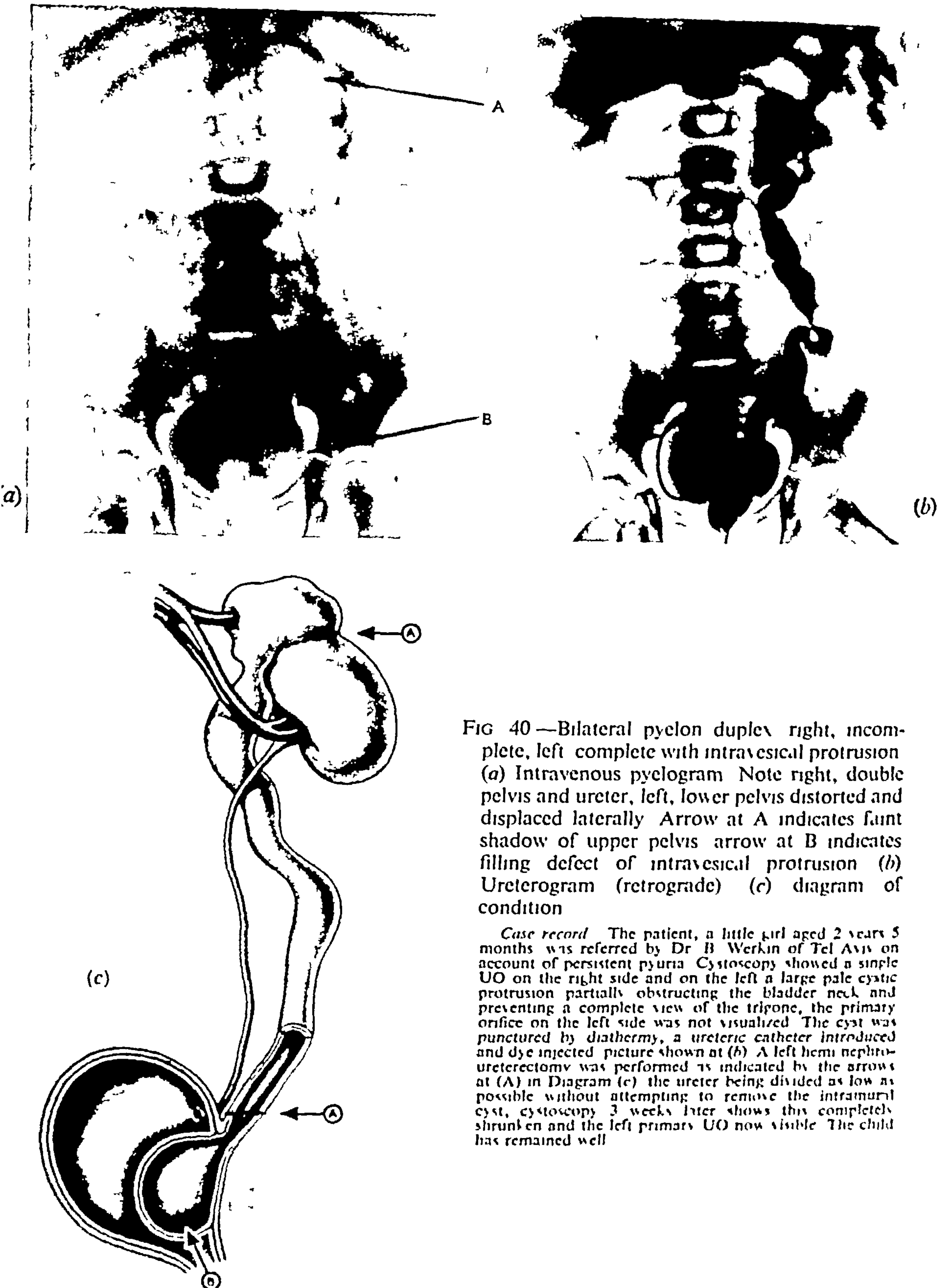


FIG 40—Bilateral pyelon duplex right, incomplete, left complete with intravesical protrusion (*a*) Intravenous pyelogram Note right, double pelvis and ureter, left, lower pelvis distorted and displaced laterally Arrow at A indicates faint shadow of upper pelvis arrow at B indicates filling defect of intravesical protrusion (*b*) Ureterogram (retrograde) (*c*) diagram of condition

Case record The patient, a little girl aged 2 years 5 months was referred by Dr B Werkin of Tel Aviv on account of persistent pyuria Cystoscopy showed a single UO on the right side and on the left a large pale cystic protrusion partially obstructing the bladder neck and preventing a complete view of the trigone, the primary orifice on the left side was not visualized The cyst was *punctured by diathermy, a ureteric catheter introduced* and dye injected picture shown at (*b*) A left hemi nephro-ureterectomy was performed as indicated by the arrows at (A) in Diagram (*c*) the ureter being divided as low as possible without attempting to remove the intramural cyst, cystoscopy 3 weeks later shows this completely shrunken and the left primary UO now visible The child has remained well



Fig. 41

Fig 41.—Ectopic ureter (right) Excretion pyelogram (30 minutes) associated bladder defect well shown (*Photograph by courtesy of Mr Debenham*)

Case record Mr Debenham kindly sends the following note. This little girl suffered from continuous day and night incontinence which was cured by removal of the right hydro-nephrotic kidney which drained through single ureter opening in the distal urethra.



Fig 42

Fig 42.—Ectopic ureter (right) Excretion pyelogram (10 minutes) showing shadow of upper pelvis (R) (*By courtesy of The British Journal of Surgery John Wright and Son.*)

This girl, aged 10 years, had suffered from typical vertical incontinence since birth. A pin-point ectopic orifice could be seen on the right side of the vestibule, but this was too small to admit a ureteric catheter Exploration of the right kidney revealed typical complete pyelon duplex. Hemi-nephro-ureterectomy cured the incontinence.

ceases at the pelvic brim, and it would appear that muscular incompetence rather than obstruction is the factor responsible In other instances the lower end of the ureter is grossly dilated, simulating a second bladder

A proportion of the cases are bilateral (*see* Fig 43)

Clinical manifestations and diagnosis

The patient is practically always a girl and the history is one of "wetting" without remission from infancy, probably chequered by infection, acute, recurring or chronic

In young children gravitation incontinence, already explained, is very characteristic, and if it can be established that the child is always dry when lying down an ectopic ureter should be strongly suspected Careful search may reveal the orifice with the efflux The ureter most commonly opens at some point in the vestibule or on the lip of the external meatus Recognition is then not difficult in a good light When the opening is in the vagina or urethra, it must be sought for under anaesthesia

Radiography

If the orifice is visualized an attempt is made to pass a ureteric catheter If this can be done, a confirmatory ureterogram can be obtained. In some cases, however, catheterization proves impossible, and in these intravenous pyelography is of the greatest diagnostic value. A double ureter and pelvis may be demonstrated or the pelvis may appear low in position with the upper segment missing; this is best seen in retrograde pictures (*see* Fig 43 (*b*))

The upper renal segment, connected with the ectopic ureter, is poor in function and concentration of the dye is therefore defective, but in Figs 42, 43 (*a*) the shadow of this segment is clearly seen detached from the main pelvic shadow

Dye tests

(*i*) The bladder is partially filled with a solution of indigo carmine, 0 4 per cent, a vulval pad is applied and the child kept up and about without micturating If the pad, though wet, is dye-free after 1–2 hours, the bladder is excluded as a source of the incontinence

(*ii*) Indigo carmine injected intravenously may be seen to colour the efflux, staining a pad blue But a delay of some hours may elapse owing to poor function of the associated renal segment and, for this reason, the test has not helped much in our cases

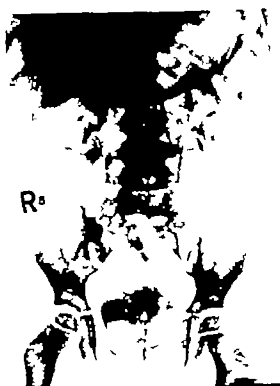
Ectopic pyelon simplex

Where a single ureter is ectopic, cystoscopy will of course show only one orifice in the bladder, and pyelography and cystography are likely to reveal associated renal and vesical anomalies

Treatment

Some authors have advocated simple ligature of the ureter at the lower end In our view the correct procedure is hemi-nephro-ureterectomy, and in the case of a single ureter, nephro-ureterectomy, provided that the contra-lateral kidney is normal.

Hemi-nephro-ureterectomy has been done in all our cases The ureter is ligated and divided as low down as possible In some instances a separate abdominal



(a)



(b)



(c)

FIG 43 —Bilateral ectopic ureter (a) Excretion pyelogram, note separate shadow of upper pelvis on both sides (b) retrograde pyelogram: lower pelves only filled (c) drawing of condition as found at operation. (*By courtesy of The British Journal of Surgery John Wright and Son*)

Girl aged 4 years Typical vertical incontinence from birth. Bilateral hemi-nephro-ureterectomy on both sides with a two-month interval. Incontinence cured

incision has been made to facilitate this, but in most cases this is hardly necessary, an extended loin incision permits of sufficient access to the pelvic portion of the ureter, where it usually narrows markedly The upper renal segment is not very clearly differentiated in every case The vessels going to it can, however, be readily isolated and when these are divided the line of demarcation is immediately clarified Haemorrhage from the kidney section is not severe and is easily controlled by mattress sutures of cat-gut passed on an atraumatic needle

The attached ureter is then passed beneath its fellow to undo the crossing and dissected clear It may be found quite narrow and attenuated at its lower end, in which case it is simply crushed and ligated as low as possible When, however, the

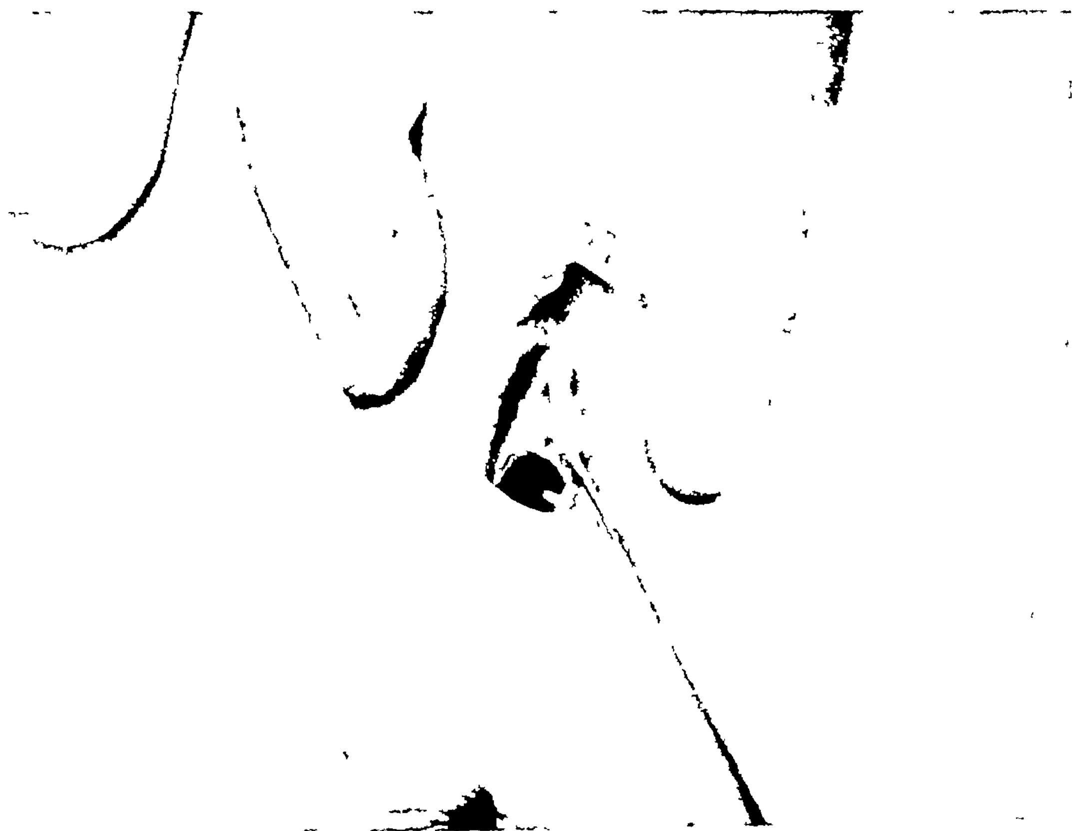


FIG 44—Ectopic ureter (left) Photograph shows orifice of an ectopic ureter in the vestibule below and to left of normal meatus

This girl, aged 6½ years had a history of recurrent febrile attacks with abdominal pain extending over 4 years and occasional incontinence by day only Treated for "pyelitis" and "enuresis"

Catheterization of both orifices as shown proved simple Urine from the normal meatus was clear that from the ectopic orifice purulent Injection of dye into the ectopic orifice outlined a left sided ectopic ureter with gross dilatation of the lower end Left hemi nephro uretereclomy in two stages cured her

terminal ureter is dilated or has formed an intravesical protrusion, something more may be required Dissection from the bladder wall is an intricate procedure and will seriously add to the severity of the operation if attempted forthwith It will usually be wiser to divide the ureter as low as possible and await events The remnant may well atrophy and further intervention be unnecessary If not, and infection persists, its removal can be carried out later with less anxiety

In one case with intravesical protrusion in which infection was the predominant feature (Fig 40) we purposed saucerizing the protrusion at a later cystotomy but the urine cleared rapidly the protrusion appeared on cystoscopy to be shrivelling and the child has remained well.

Triple ureter and diverticula

Triplication

More complex multiplicities of the ureter are possible and various appearances may result The commonest of these very rare anomalies is that in which 3 ureters are present Most often these stem off from a single original ureteric bud and there is therefore only one opening into the bladder (trifid ureter)

The condition is in fact an incomplete pyelon duplex with an additional bud. Very rare instances are on record of still more buds resulting in further multiplicity of the ureter



FIG 45—Ureteral triplication. Retrograde pyelogram.

Infant girl aged 9 months with chronic pyuria. Right upper tract normal Trifid ureter on left side (single vesical orifice) Left nephro-ureterectomy Cure. (*By courtesy British Journal of Surgery John Wright and Sons.*)

Triplication may also occur in association with a complete pyelon duplex, when there will be at least 2 openings into the bladder and quite possibly 3 (complete triple ureter) In some cases localized sacculations indicate a fundamental error of development in addition to the supernumary budding.

Clinical

Such complexity of the ureteral stem renders it particularly liable to obstruction and infection and the presenting symptoms will be pain pyuria and possibly

haematuria. The radiographic picture is likely to be bizarre but rarely diagnostic Withycombe (1950) recorded an interesting case in an adult in which the uretero-gram demonstrated a high trifurcation. Usually the diagnosis will only emerge at operation and nephro-ureterectomy be the only possible course

One instance of triplication has occurred in our series This was recorded by Smith (1946) (*see* Fig 45) The two upper ureters were dilated and tortuous, the lowest normal One orifice only was seen in the bladder and the case was one of trifid ureter

Diverticula

Diverticulum of the ureter has only been encountered as an incidental finding (*see* Fig 53), but cases are reported in which enormous sacs have been formed, these may contain stones and obstruct the ureter Diagnosis in such cases has usually only been made at operation, though in some it was suspected from the retrograde

Ureterocele, megaureter and congenital stricture are discussed elsewhere.

CHAPTER 8

URINARY OBSTRUCTION—VESICAL RETENTION

ARREST (retention) or retardation (stasis) of the normal urinary flow at various levels is so frequent in the young that a large part of children s urology is concerned with its elucidation and treatment The manifestations are those of dilatation of varying type and degree, almost inevitably bedevilled sooner or later by infection While the outstanding dilatation as seen in the individual case may appear local in character for example hydro-nephrosis, megaureter or vesical retention, the close integration of the tract as a whole must be borne in mind if a true under standing of the pathological evolution of many of these obstructive lesions is to be achieved.

Site of obstruction

This may be (i) in the infra vesical zone (bladder neck or urethra) (ii) at the uretero-vesical level (iii) at the pelvi ureteric level

The obstruction may be complete or incomplete and in this respect, infection inevitably introduces an element of variability The presenting clinical features will vary accordingly but broadly speaking they will conform respectively to those of 1 vesical retention 2, megaureter and 3 hydro-nephrosis.

While such is the broad outline the problem in practice is often less simple and the clinical picture less definitive One thing very much leads to another in the urinary tract and the condition as we happen to see it at the moment may well be that of multiple complex dilatation in which the initiating lesion may be obscured by the sequelae or may even indeed have resolved, leaving these as the sole mani festation Such complications make a right understanding of the pathogenesis of many of these cases a matter of great difficulty and decisions as to treatment by no means easy Thus it may not infrequently happen that the secondary *lesion* demands primary consideration in treatment, for example secondary obstructive hydro-nephrosis due to inflammatory adhesions (*see* page 154)

Nature of obstruction

Along with determination of the site or sites of obstruction and an assessment of the relative clinical significance must go consideration of the nature of the lesion This may be (i) organic some well defined structural abnormality congenital or acquired (ii) what we must still term functional (achalasia) where no such structural abnormality is demonstrable

Under this second heading falls a group of cases which is of considerable practical importance in children because the aetiology is still conjectural and the treatment therefore uncertain Our experience leads us to believe that, with advancing knowledge an increasing number of the cases now classified as achalasia will be found in fact to have an organic origin

To summarize, therefore, children suffering from "obstructions" in the urinary tract will present to us in varied clinical settings, in which micturition difficulties, pyuria, haematuria, pain, "tumour", and grave defects of health and bodily growth due to renal failure may all feature

The manifestations may give a clue to the level of obstruction, but it is for the surgeon to establish as conclusively as may be its exact site and nature, and to adjudge the urological findings in relation to renal function and general health as a preliminary to effective treatment

Infra-vesical obstruction (retention)

Infra-vesical obstruction in children may be due to a considerable variety of lesions, both within and without the urinary tract, which differ widely from one another both in the degree of obstruction and the rapidity of their onset At one extreme are the minor causes of acute retention which can be relieved as soon as diagnosed at the other the serious anomalies of the urethra and bladder neck, which insidiously, but irreversibly, destroy the renal function Although the modern methods of investigation, particularly urethroscopy and cysto-urethrography, have elucidated many of the problems of these cases, the cause in some is still obscure either because at post-mortem examination no lesion can be found below the dilated segment, or because spontaneous cure appears to have followed simple drainage of the bladder

Acute retention

Acute retention, with complete inability to micturate and painful distension of the bladder, is a clinical picture which can scarcely be mistaken or overlooked It may of course be the culmination of a slowly increasing obstruction, though this is not so common in children as in the prostatic hypertrophy of old age, and upper tract dilatation will always betray the chronic nature of the disease

Acute retention may be due to quite minor causes An infant with a meatal ulcer is apt to hold his water until the very last minute and then cry as the stream of urine tears the scab from the surface of the ulcer, and this pain often results in acute retention We have had several cases in which a blockage of the external meatus has been caused by a plug of sulphamerazine crystals, and at times it seems that the irritation of the passage of oxalate crystals is sufficient to cause a child a temporary retention A stone impacted in the posterior urethra may cause a complete stoppage, and the following two cases provide interesting examples of extra-urinary conditions causing acute retention

Acute retention in pubic osteomyelitis

Boy, aged 4 years, admitted with supra-pubic swelling and acute retention Attempts to pass urine caused severe pain and produced only an occasional dribble with prompt stoppage On examination, a very tender mass could be localized in front and slightly to the right of the bladder, which was tightly distended Under anaesthesia a gum elastic catheter passed easily and drew off 5 ounces clear urine Radiological examinations were negative W B C 11 000

During the succeeding days the lump became less tender and more localized Micturition gradually became normal Eventually a small abscess formed and was incised (*Staph aureus*) Later skiagrams showed a focus in the os pubis Recovery uneventful

Pyocolpos producing acute retention

This condition is occasionally seen in baby girls. A typical case (D.S aged 4 months) presented with an abdominal tumour and a bulging hymen together with complete retention. Withdrawal of the urine by catheter was found to leave the abdominal lump and the vulval bulge much as before The hymen was freely incised and the vaginal contents (pus and altered blood) evacuated. There was no further trouble

Chronic retention

In cases of chronic retention the cause is not always immediately obvious, and in some, treatment of the retention must precede diagnosis of the cause. Despite individual variations, the general effects of chronic urinary obstruction are similar in all cases and it is convenient to describe first the pathology and clinical picture of chronic obstruction in general and then the particular lesions with their distinctive features

The effects of obstruction

Pathology

The bladder at first responds to obstruction of its outlet by muscular hypertrophy and in this way may be able to compensate for a partial obstruction In children it is often found that this hypertrophy is sufficient to empty the bladder completely but the back pressure effects are borne upon the upper urinary tract Sooner or later however the bladder contraction fails and after micturition a residue is left which gradually increases until vesical distension is evident The hypertrophy may show cystoscopically as trabeculation and as a prominent inter ureteric bar Yet a perfectly smooth surface is occasionally found upon an extremely thick wall and the absence of trabeculation should not preclude the possibility of an obstruction

Diverticula though rare in childhood have been found in association with the common forms of obstruction and their presence is apt to accentuate the symptoms Most diverticula are only congenital in the sense that they are formed at the site of a congenital weakness in the bladder wall and the pouch is only developed under conditions of increased infra vesical pressure such as results from obstruction. It is possible that there are a few cases of truly congenital diverticula but it should always be assumed that obstruction is present. The characteristic site is postero-lateral to the ureteric orifice, but in children it is common to find that the ureter actually opens within the sac the inter ureteric bar can often be traced over the rim and a normal slit like orifice is found a few millimetres inside These ureters are apt to be obstructed by the pressure of the sac and by fibrosis around its wall in which they become involved In the following case occurring in infancy a diverticulum burrowed downwards and lay directly behind the posterior urethra, which it compressed causing acute retention

Baby J P Male. Aged 9 months.

Was admitted under the care of Mr Denis Browne as a suspected intussusception. The abdominal pains settled down but increasing distension of the bladder was noticed. 18¾ ounces urine were withdrawn by catheter but retention recurred and continuous catheterization was necessary Blood urea 39 mg. per 100 ml. Intravenous pyelogram showed dilation of both ureters of moderate severity kidneys of good function. Per rectum a firm, slightly tender swelling was found in the mid line below the bladder in the position of the posterior urethra.

The findings suggested an infected cyst of the utriculus. The child's general condition improved, normal micturition was resumed and he was discharged.

Three months later he was re-admitted with recurrence of retention. The swelling could still be felt per rectum. Cysto-urethroscopy showed a severe basal cystitis and a bulging trigone. The ureteric orifices could not be visualized because of the general oedema. The posterior urethra appeared normal.

It was decided to open the bladder and approach the retro-vesical swelling, if necessary through the trigone. On opening the bladder, a diverticulum was found at the left ureteric orifice. This extended downwards behind the bladder into the retro-urethral area. The diverticulum was excised and a wedge resected from the bladder neck.

The infant made a straightforward recovery and normal micturition was resumed in 10 days. He has remained well.

The ureters become dilated and their muscle hypertrophied. At first the dilatation tapers off towards the bladder, but later the wall appears thin and flabby, the ureter is tortuous, and distended pouches may be formed at the lower end. The ureteric orifice is often entirely normal, dilatation of the intramural ureter being prevented by the pressure of the hypertrophied detrusor, this is particularly the case where the bladder is "compensating" at the expense of upper tract dilatation. On the other hand the orifice may be relaxed and allow a free reflux. It may even appear cystoscopically as a wide black hole, though it retains its characteristic obliquity and may be likened to a railway tunnel (*see* Fig 68). The reflux increases the ureteric dilatation but for a time this dilatation itself acts as a buffer to protect the kidney and a comparatively normal pelvis may surmount a huge ureter (Defence in depth *see* Ureteral Reflux, Appendix 1).

In cases where the obstruction has been present since early foetal life, the kidneys are often found to be small and cystic, the cysts being apparently formed within the parenchyma without gross communication with the pelvis. Such kidneys are incapable of supporting life for long and are found only in the stillborn or neonatal fatalities. Obstruction of later onset or slower progress produces typical hydro-nephrosis, as in adult life, though the kidney may appear and feel lobulated because of the distended calyces, so that cases in which vesical distension is not obvious may be mistaken for polycystic disease.

The effect of back pressure upon the nephrons is to produce a slow deterioration of function with the formation of a dilute urine and consequent polyuria. A slow but steady rise of the blood urea, which may reach extreme figures, is a feature of the chronic cases, and the persistent uraemia is responsible for underdevelopment and dwarfism. The effects upon the skeleton of the acidosis which accompanies uraemia and the production of renal rickets have been discussed in Chapter 4.

Clinical picture

The symptoms of chronic urinary obstruction may be associated with disorders of micturition or with the biochemical disturbance of renal failure or with both. The urinary stream may be poor, the child having to strain and taking a long time to complete the act, but an apparently normal stream does not rule out the possibility of obstruction since, as we have seen, the bladder hypertrophy may compensate, though at the expense of the upper tract. Occasionally parents will mistake the slight grunt of satisfaction given by a normal infant on passing urine

for a pathological straining, and it is unwise to place much reliance upon a simple history of difficulty Whenever possible the child should be watched passing urine.

As in the prostatism of old age there may be frequency nocturnal enuresis, or a continuous dribbling incontinence resulting from an over-distended bladder Palpable distension of the bladder after micturition is obviously indicative of an obstruction yet in infants it is often difficult to be sure that the act of micturition has in fact been completed Moreover a bladder containing only a small quantity of urine may be easily palpable in the child particularly if there is a mass of faeces in the rectum

Palpable enlargement of the kidney is not infrequent in severe obstruction

Infection always precipitates a serious deterioration of function a slight infection may cause acute retention where there has been only a partial obstruction or a severe infection may overwhelm the child so rapidly that the obstructive element is not suspected until post mortem examination *Infra vesical obstructions* are peculiarly liable to infection following instrumentation even where the most stringent precautions have been observed and where it can be avoided the passage of catheters or cystoscopes should be postponed until all preparations to deal directly with the causal lesion are complete The investigation of cases of recurrent urinary infection only rarely reveals a case of infra vesical obstruction much more often upper tract lesions are found

Stone formation is not a common accompaniment of these cases in contrast to enlargement of the prostate perhaps the dilute urine so often seen in the child with lower tract obstruction does not favour the deposition of crystals.

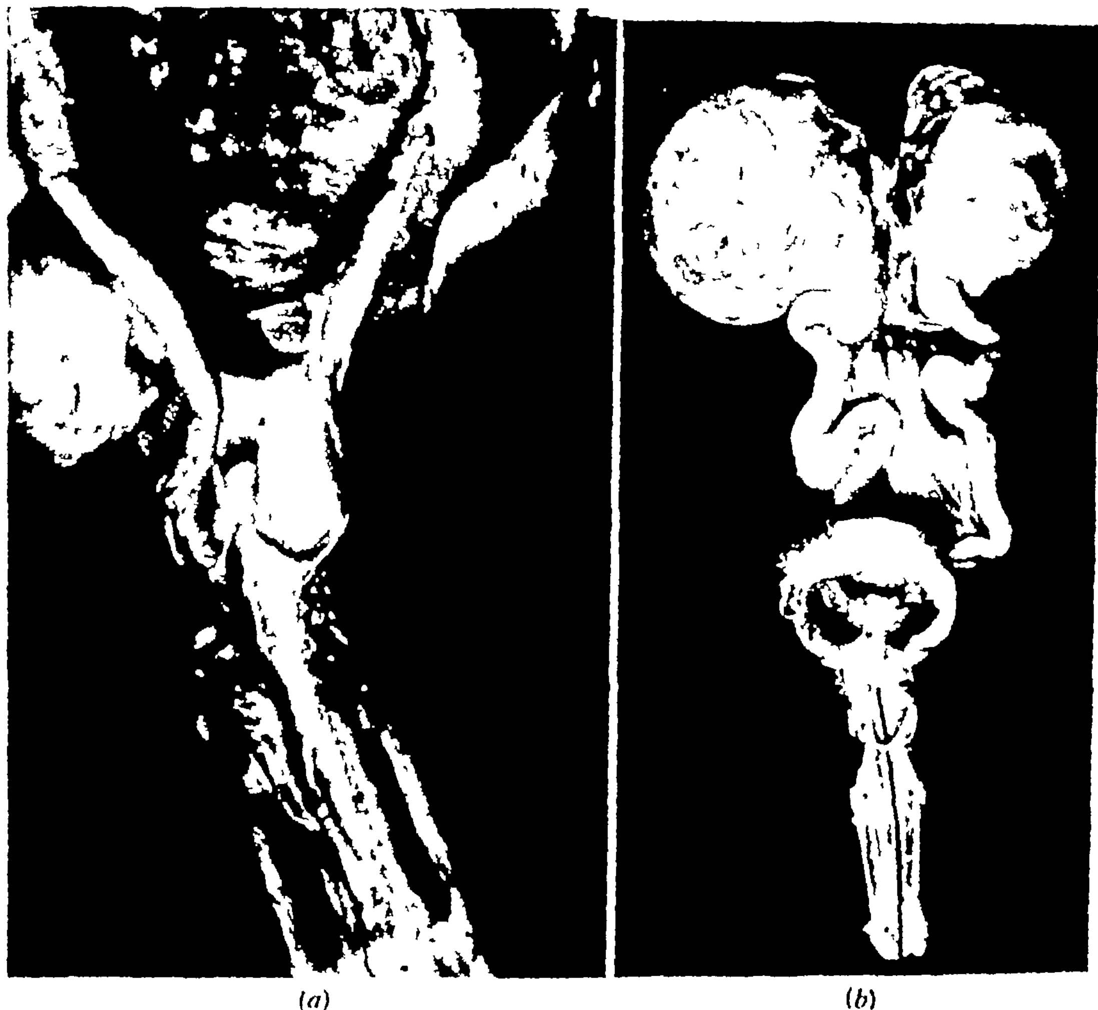
THE CAUSES OF CHRONIC OBSTRUCTION IN THE MALE

Congenital valves in the posterior urethra

There can be no doubt that valve like mucosal flaps in the posterior urethra are an important cause of urinary obstruction in the infant, and account for most of the fatal cases during the first year of life Careful dissection is needed to display them and in earlier post mortem records they have often been overlooked The great majority of cases correspond to Young s Type I (Young, Frontz and Baldwin 1919) and are situated below the verumontanum Often bicuspid, their free edges may be prolonged upwards as prominent ridges on the floor of the urethra or they may be fused together in the mid line, forming an almost complete curtain and leaving only a small median aperture anteriorly Their general similarity to the normal submontanal folds has already been pointed out (Chapter 6) and it is believed that they represent the persistence and adhesion of normal large folds found in the foetus The interval between the folds and the verumontanum varies from case to case sometimes the obstruction is well down in the membranous urethra (*see* Fig. 46 (*a*) and (*b*)) The ballooning effect of the urinary stream directed into the pouch between the fold and the lateral urethral wall is probably respon sible for the increasing obstruction It must, however be realized that the hold up of the urine during foetal life may have largely destroyed the renal substance by the time of birth

We have not encountered a definite case of Young s Type II or III nor have we been able to find a museum specimen illustrating such a case Type II is described as having the folds running upwards from the verumontanum towards

the bladder neck, and it should be noted that in the ordinary submontanal cases there are often prominent fibrous ridges in this situation, though they are not the obstructive element Type III is an "iris" diaphragm occurring at any level without reference to the verumontanum, and since this form alone is apt to be destroyed by the passage of instruments *per urethram*, these cases are likely to be confused with "congenital stricture"



(*a*) (*b*)

FIG 46—Specimens illustrating urethral valves (Museum, Hospital for Sick Children, Great Ormond Street) (*a*) From a baby aged 7 months death from infection and renal failure, (*b*) from a baby aged 5 weeks death from renal failure (note the fused kidney)

Hypertrophy of the bladder and changes in the upper urinary tract are always well marked in the valve cases Dilatation of the posterior urethra, above the obstruction, is always obvious and may be extreme The soft compressible mass produced by this dilatation may occasionally be felt at rectal examination The bladder neck partakes in the general hypertrophy of the detrusor muscle, and appears as a well marked constriction, which may shut off the bladder from the urethra This fact is of some importance, since in the simple cystogram the urethra may not fill and the obstruction might be supposed to be at the bladder

eck a micturating cysto-urethrogram will reveal the true state of affairs (*see* igs 47–48)

At urethroscopy the valvular folds themselves are hard to identify because the tream of irrigating fluid tends to flatten them against the lateral urethral wall rominent ridges on either side of the crista urethralis dilatation of the urethra nd a patulous internal sphincter are suggestive urethroscopic signs



Fig 47—Valvular obstruction Micturating cysto-urethrogram showing gross dilatation of proximal urethra.

The baby aged 6 months, suffered with difficult micturition and straining from birth. Pyuria had been present since the age of one month. Blood urea [illegible] mg. per 100 ml. Right kidney palpable. Bladder could not be felt. Cystotomy and retrograde dilatation of urethra by sounds was performed. Satisfactory micturition was ultimately re-established, but the urinary infection could not be completely controlled. He ultimately died 2 years later from the effects of renal damage.

Marion s disease: bladder-neck obstruction

Although many surgeons from Guthrie (1834) onwards, have described obstruction at the bladder neck due to spasm muscular commissure fibrous bar or neuro-muscular dysfunction Marion (1933) was the first to give a comprehensive account of the condition of its pathology and of its onset in early life Thus since the aetiology remains obscure it is convenient to speak of Marion s disease

The condition has much in common with achalasia elsewhere in the body and in the infant and child it may be hard to identify any definite pathology at the site of the obstruction. the whole detrusor muscle is hypertrophied and the bladder neck partakes in that hypertrophy At operation the firm closure of the internal sphincter and its resistance to digital dilatation is characteristic, and a simple division of the fibres of the posterior lip often lays the bladder neck wide open. The hypertrophy of the muscle at the internal sphincter may often be appreciated cystoscopically as a bar and in the course of time fibrous infiltration may render the muscle sclerotic.

Diverticula are more commonly found in association with Marion's disease than with other causes of obstruction These diverticula have sometimes been regarded as congenital pouches, and the muscular hypertrophy of the bladder attributed to the hindrance to micturition consequent upon their presence The repeated observation that simple excision of the sac, without interference with the bladder neck, is frequently followed by the appearance of other signs of obstruction confirms the view that the diverticula are in fact secondary (Badenoch, 1949)



FIG 48 —*Valvular obstruction Micturating cysto-urethrogram (A P and lateral views)*

Case record Male, aged 9 years, referred on account of frequency, attacks of urinary infection and abdominal pain The child's mother had noticed that he had been very thirsty since the age of 9 months and he had had attacks of screaming on micturition as a baby On examination he was considerably under weight, the kidneys were not palpable but the bladder could be felt just above the symphysis after micturition The urine was sterile at the time of examination Blood urea was 125 mg. per 100 ml Residual urine 6 ounces Intravenous pyelography showed very poor function and faint shadows indicated gross dilatation of both ureters This was confirmed by retrograde pyelograms The voiding cysto-urethrogram showed an irregular outline to the bladder, no reflux and marked dilatation of the posterior urethra, with the outline of folds beneath the verumontanum

At retro-pubic urethrotomy two mucosal valves were demonstrated and destroyed by diathermy The wound was closed with supra-pubic drainage Convalescence was complicated by a febrile attack with severe loin pain, which cleared up on streptomycin On discharge he was passing a good stream, there was no residual and the urine was sterile Blood urea was 82 mg per 100 ml A year later his general health was moderately satisfactory there was no residual urine but he was suffering from some incontinence both by day and night The urine was of very low specific gravity and the blood urea had still not fallen to normal

Clinically we have not found Marion's disease common in early infancy Few of our patients have been under 3 years of age The pre-operative diagnosis has been established chiefly by the elimination of other causes, in particular the demonstration of a normal posterior urethra by cysto-urethrography The cystoscopic sign of a collar at the bladder neck may not always be identifiable

It must be emphasized that the true Marion's disease is not of demonstrable neurological origin The typical neurogenic bladder seen in association with spina

bifida is recognizable not only from the accompanying nervous system abnormalities, but by the ease with which urine can be expressed by manual pressure upon the abdomen by the early vesico-ureteral reflux, and often by filling of the posterior urethra In our experience trabeculation and the bar at the bladder neck are often more prominent in the neurogenic bladder than in Marion s disease. There is no evidence that a partial nerve lesion can be responsible for the true Marion picture and a purely radiological diagnosis of spina bifida occulta is of no consequence in this connection (*see* Figs. 49 and 50 (*a*) and (*b*))

The intravesical causes

A mild chronic retention may result from the presence of a ureterocele which impinges upon the internal meatus, or more frequently of a cystic intravesical protrusion of the lower end of an ectopic ureter since the bladder neck itself is often involved in these cases (*see* Fig. 39) Rhabdomyosarcoma of the bladder presents as retention of urine since the fleshy lobules tend to plug the internal meatus where the growth arises within the prostate, retention is also a feature because of direct pressure upon the urethra (*see* Chapter 13)

Cases have been described (Learmonth and Watkins, 1935) in which a curtain of vesical mucosa, arising from the trigone, appeared to obstruct the bladder neck It is difficult to be sure that such cases are not examples of Marion s disease, associated with redundant oedematous mucosa The only case in our experience which at first appeared to correspond with the description of a trigonal curtain proved to have a double ureter with cystic protrusion of the lower end (*see* Appendix III case 1)

Stricture

Congenital stenosis of the external meatus may be observed on occasion, especially in association with glandal hypospadias, but it is rarely a cause of more than a temporary retention A rare fatal instance however is illustrated in Fig. 51 Phimosis cannot be regarded as an important cause of dilatation of the urinary tract

Congenital fibrous stricture of the bulbo-membranous urethra has been described in the infant (Schmidt, 1924) but it is generally admitted to be a rare condition The diagnosis congenital stricture is more often made in adolescents in whom no history of infection can be obtained and in whom a single thorough dilatation is not followed by recurrence This type of stricture is to be regarded as a mucosal diaphragm analogous to the obstructive valves rather than a long fibrous constriction the following case illustrates this condition

C W Male. Aged 5½ months

Admitted under Dr Paterson because of continuous dribbling incontinence with a bladder distended up to the umbilicus. Blood urea 32 mg. per 100 *ml* and urine sterile.

At operation attempts were made to pass instruments into the bladder but they were held up in the membranous urethra and a supra pubic cystostomy was performed. Some days later it was possible to pass a sound and the urethra was gradually stretched by a series of dilatations under anaesthesia.

In the following year he was re-admitted with a urinary infection but his temperature settled quickly and he was dry day and night. He had no further urinary symptoms but



Fig 49.—Bladder-neck obstruction (Marion) Male aged 3 years Micturating cysto-urethrogram Note "fir-cone" bladder and some dilatation of proximal urethra

Case record This boy was admitted under Dr Lightwood with the tentative diagnosis of polycystic kidneys, and suffering from wasting, apathy, urinary incontinence and frequency He had a history that during the first week of life he had a laparotomy and the kidneys were felt to be enlarged at that time

On examination he was very drowsy and wasted, and was running a moderate pyrexia Both kidneys were enlarged and palpable and the bladder was distended up to the umbilicus Urine was heavily infected with *B proteus* and his blood urea was 270 mg per 100 ml Haemoglobin 47 per cent

A bilateral nephrostomy was performed as an emergency operation and he was given a blood transfusion Nephrostomy drainage was continued for 4 weeks, during which time his blood urea dropped to 78 and the urea concentration of his urine rose His general condition improved beyond recognition A cystogram showed a reflux to dilated ureters and renal pelves, and during a forceful bladder contraction the posterior urethra appeared to be somewhat dilated The bladder was trabeculated

Some days later the bladder and posterior urethra were opened by retro pubic approach The posterior lip of the bladder neck was hypertrophied and sclerotic. No valves could be found in the posterior urethra A wedge of tissue was removed from the bladder neck and the wound closed with a supra pubic drain Normal micturition was re-established in 6 weeks by which time his general condition was very greatly improved The child has remained extremely well



(*a*)

Fig 50.—Bladder-neck obstruction (Marion) Male aged 8 years. (*a*) Voiding cysto-urethrogram. Proximal urethra not dilated. No reflux (*b*) Intravenous pyelogram. right, hydro-nephrosis and megaureter left, no secretion (retrograde confirmed the presence of a similar though more advanced, degree of dilatation on this side)

Case record This boy was referred by Dr Evans because of recurrent urinary infection and continual dribbling incontinence. He had first had an attack diagnosed as "pyelitis" at the age of 5 months.

On examination he was somewhat underdeveloped, both kidneys were palpable but not grossly enlarged, and after micturition the bladder remained 3 inches above the symphysis. The urine was moderately infected with *B. procyaneus* and the blood urea was 28 mg. per 100 ml.

Cystoscopy revealed a trabeculated bladder with normal ureteric orifices. A micturating cysto-urethrogram showed normal posterior urethra and no reflux into the ureters. Intravenous pyelogram revealed no function on the left side, hydro-nephrosis and hydro-ureter on the right. The bladder was opened supra-pubically and wedge cut from the lip of the internal meatus, which was hypertrophied and sclerotic. The bladder was closed with a supra-pubic drain. The wound closed rapidly after the removal of the supra-pubic tube, and micturition was re-established. His pyocyaneus infection remained and he had a tendency to incontinence.

Nine months later he was well, the urine clear and he was dry by night although occasionally wet by day. There was no residual



(*b*)



FIG 51 —Photograph of specimen from a male infant aged 2 weeks, to illustrate fatal retention resulting from extreme stenosis of the external urinary meatus. The kidneys were cystic Note the urethral dilatation

Case record The infant had acute retention from birth and was admitted with the story that all previous attempts to pass a catheter had failed The external meatus had to be incised before a ureteric catheter could be introduced He was obviously in an advanced stage of renal failure and despite continuous catheter drainage and transfusions death ensued 24 hours after admission



G 52 —Urethrogram showing bifid penile urethra Male aged 12 years The anomaly caused valvular obstruction to micturition (*By courtesy of Edward Arnold and Co*)

was re-investigated 5 years after the initial operation The bladder was still trabeculated and the posterior urethra slightly dilated There was however no significant residual and micturition appeared to be normal There was well marked dilatation of the left kidney and ureter

The passage of instruments on very young infants is occasionally difficult, particularly if sounds with the full adult curve are employed, and the label stricture should not be attached to the case unless definite gripping of the instrument can be felt

Traumatic stricture of the anterior urethra is not very uncommon It may result from external violence (falls astride a fence and similar mishaps to which small boys are liable) or from injudicious urological instrumentation Indwelling catheters are occasional causes of this complication

Inflammatory strictures, due to gonorrhoea in older children have been described (Campbell, 1937) but we have not observed them

Cysts and diverticula of the urethra

Cysts arising in the verumontanum, some of them clearly dilatations of the utriculus masculinus may on rare occasions be responsible for urinary obstruction Wainwright (1951) has recorded a case found at *post mortem* in an infant who had died of uraemia Badenoch (1949) has mentioned a case in which a cyst of the seminal vesicle was the obstructive factor One instance of a seminal vesicalular cyst has been encountered by us and this caused no urinary symptoms but presented as a large cystic abdominal swelling rising out of the pelvis.

Diverticula of the anterior urethra, which fill with each act of micturition may form a valvular obstruction though they are likely to be recognized before the effects of retention are very far advanced (Johnson 1923)

In one case, an accessory urethral canal in the penis was responsible for severe chronic retention the point of union of the two passages having a valvular effect.

K G Male Aged 12 years.

History 2 years micturition difficulties and infection On examination bladder distended right kidney palpable urine sterile residual urine 13 ounces Intravenous pyelogram showed bilateral hydro-nephrosis and megaureter poor secretion from right Cystogram revealed gross bilateral reflux and the urethrogram a bifid urethra (*see* Fig. 52) The external orifice of the dorsal segment was minute, and it appeared that obstruction to micturition resulted from ballooning of the upper urethral segment. The dividing valve was cut through and recovery was uneventful

Congenital hypertrophy of the verumontanum

Bugbee (1923) first described clinical and post mortem cases in which a considerable enlargement of the verumontanum blocked the posterior urethra, causing all the changes in the upper urinary tract commonly seen in cases of congenital valves in the posterior urethra. There can be no doubt that there is a considerable variation in the size of the normal verumontanum, and even where it seems large, it is difficult to be sure that bladder neck disease of the Marion type is not the cause of retention. In recent papers on the subject of retention it does not appear that this diagnosis is often made, but in one of our cases it was felt that the verumontanum was the responsible factor (*see* Fig 53)



Fig 53

FIG 53 —Infra-vesical obstruction attributed to verumontanal hypertrophy Excretion pyelogram from male aged 7 years Note the dilated proximal urethra and diverticulum of the right ureter

This boy was investigated on account of enuresis and diurnal frequency The urine was sterile and the blood urea 27 mg per 100 ml The cystogram showed trabeculation of the bladder, a funnel-neck deformity and reflux into the right ureter Residual urine 1 ounce. Cystoscopy and retrograde pyelogram confirmed the above findings Urethroscopy showed an enlarged and inflamed verumontanum This was treated by massage with a finger in the rectum and a bougie in the urethra Subsequently the bladder was opened and sounds passed in a retrograde direction but no obstruction could be felt

Six years later there were no urinary symptoms An intravenous pyelogram showed a normal upper urinary tract, the enuresis, which had been troublesome for some years, had finally cleared up during psychological treatment (*By courtesy of Edward Arnold and Co*)

FIG 54 —Atresia Ani Urethralis Voiding cysto-urethrogram Boy aged 5½ years Note the marked kinking of the proximal urethra

During operation for restoration of an imperforate anus, both ureters were noted to be considerably dilated Subsequent cystography showed a kinked posterior urethra During the following 2 years he gained some slight control over his bowel actions but urinary infection was troublesome Finally the recto-urethral fistula reopened spontaneously, although it did not allow a great deal of leakage Re-investigation showed a somewhat greater degree of dilatation of the ureters, a trabeculated bladder though no residual urine and an elongated posterior urethra which was dilated down as far as the region of the bowel where there was a stricture



Fig 54

Atresia and urethralis

Many of the cases of imperforate anus in the male have a fistula, or a fibrous connection with the posterior urethra usually in the region immediately below the verumontanum (*see* Chapter 6) This fistula is difficult to eliminate and apt to recur after operation it may be responsible for recurrent urinary infection A serious chronic retention of urine is also a feature of some cases and may be responsible for deterioration of the child s condition after the bowel obstruction has been relieved A study of museum specimens suggests that urinary obstruction may be present even where the fistula and lower bowel have not been interfered with and urethrograms make it clear that the urethra is kinked at the site of the fistula, being drawn upwards and backwards by the contraction of the bowel (*see* Fig 54) Our clinical experience suggests, however that the obstructive element is more severe after attempted restoration of the rectum After such operations the urethra may be elongated and displaced from the mid line it may be dilated and 2 cases showed definite relaxation of the bladder neck suggesting some damage to the nervi erigentes at the time of the rectal operation The scar in the perineum may deform the membranous urethra, and one case seemed to have a stricture in this region

Absent abdominal muscles

There is an interesting and obscure group of male children in whom the lower abdominal muscles are either entirely absent or reduced to a fibrous sheet. In all recorded cases there has been some dilatation of the urinary tract, and in the most carefully examined cases this dilatation appears to have extended down as far as the posterior urethra The type of obstruction at this situation has not been adequately described, but does not seem to correspond with the typical congenital valves Some authors (for example Eagle and Barrett 1950) take the view that obstruction may occur at varying levels in the urinary tract, but as has been pointed out, it is easy to be misled concerning the site of obstruction unless the fullest investigations are carried out. It has been supposed that the absence of pressure within the abdomen, consequent upon the muscular deficiency is the cause of the urinary tract dilatation but the characteristic distribution of the defect suggests rather that vesical distension in foetal life has interfered with the development of the muscles The bladder usually remains adherent to the umbilicus, and the testes are intra abdominal. Death is frequently due to renal failure in early life but the condition is not incompatible with survival beyond childhood.

Treatment of the urinary obstruction must be guided by the findings in any particular case in a personally observed instance the bladder although distended emptied without residual and no treatment was undertaken (*see* Fig 55 (*a*) and (*b*))

THE CAUSES OF CHRONIC OBSTRUCTION IN THE FEMALE

Obstructions to the lower urinary tract are naturally not so common in girls as in boys, and in them the causes are often obscure The intravesical causes are of course, the same, and a ureterocele may even prolapse through the entire length of the female urethra A stenosis of the external meatus is occasionally seen as a

cause of mild retention, otherwise strictures do not occur The bla obstructions may occur in girls as in boys, though in the former we h that a wedge resection of the internal meatus has often disappointing

In some cases, in which there is no evidence of a neurological lesion dilatation of the upper part of the urethra, which fills in a simple cysto exact site of the narrowing cannot at present be described in anatomical



(*a*) (*b*)

FIG 55—Vesico-urethral dilatation associated with congenital agenesis of abdc muscle Male aged 1 year (under the care of Mr Charles Donald) (*a*) Photogra the child, (*b*) cysto-urethrogram Note urachal diverticulum and dilated proximal ur

This child had been wearing a supporting belt from the first few months of life Following an at haematuria and urinary infection, urological investigation was undertaken Both ureters were consi dilated and the bladder was of very large capacity with a saccular dilatation reaching up to the um The posterior urethra was also dilated There was no residual urine Infection cleared by chemothe

it is uncertain if any mucosal folds similar to the valve in the male ure occur. Morphologically, of course, the female urethra corresponds onl supra-montanal urethra of the male, and the embryological explanation a for the submontanal valve would be invalid (*see* Figs 56 and 57).

TRANSIENT OBSTRUCTION

There is a small, and doubtless heterogeneous, group of cases in which urinary obstruction, having persisted long enough to cause upper tract dil and perhaps even to require catheter or supra-pubic drainage, appears to and allow the re-establishment of normal micturition We suggest that som



Fig. 56

FIG 56.—Cystogram in infra vesical obstruction in a girl aged 5 years showing trabeculated bladder with reflux into grossly dilated ureters and kidneys.

Child referred by Dr Wyllie with severe frequency of micturition (half-hourly by day) and a long history of constipation. On examination she was under developed, and a distended bladder was palpable after micturition. Residual urine 5 ounces (infected *B. coli*). Blood urea 108 mg. per 100 ml.

Cystotomy Wedge resection of bladder neck. Following operation her general condition improved, BU fell to 71 and after 4 months to 57 The suprapubic track, however would not heal despite repeated urethral dilatations and excision of the fistula She finally died 7 months after operation, the immediate cause of death not being obvious.

FIG. 57.—Infra vesical obstruction in a girl aged 4 years. Cystogram showing very large bladder with filling of the proximal urethra.

This child first attended at the age of 6 months on account of obstinate constipation. In the following year it was noticed that her urine was infected and the bladder distended. She was usually dry by night but wet by day Residual urine 11 ounces (infected *B. coli*). Intravenous pyelogram showed no abnormality of the upper urinary tract. On cystoscopy the bladder was trabeculated but there was no evident bladder neck obstruction. N neurological signs.

Cystotomy Wedge resection of bladder neck. Fistula closed quickly and the symptoms abated, the child being dry by day and night Urinary infection has been kept under control by chemotherapy but small residual (2–4 ounces) is still usually present. After 2 years her general health remains good and the upper tract undamaged



Fig. 57



Fig 58

Fig 58—Transient infra-vesical obstruction Infant girl aged 6 months Cystogram (dye introduced through cystotomy wound) showing reflux and filling of perinephric abscess cavity (right) (*By courtesy of Dr Gairdner and Mr Vernon Pennell*

Fig 59—Transient infra vesical obstruction Cystogram from male infant aged 18 months, showing bilateral reflux and dilatation of upper tract

This infant was admitted in great pain with acute retention and a distended bladder from which 16 ounces of clear urine were withdrawn by catheter Blood urea 219 mg per 100 ml (after 2 days catheter drainage fell to 106) Cystoscope passed with ease and revealed trabeculation and oedema of the base Retention continued and a cystotomy was performed Retrograde passage of sounds met with no apparent obstruction in the urethra Following the cystotomy the BU fell rapidly, normal micturition was re-established and at 4 years the child was symptom-free and his urine sterile Blood urea 47 mg per 100 ml Re-investigation showed that the dilatation of the upper tract had persisted but the bladder and posterior urethra showed no abnormality

This child may have had congenital urethral valves which were destroyed by the passage of sounds, but there was no definite evidence for this by urethroscopy or otherwise The persistence of upper tract dilatation, despite restoration of normal bladder function, is an important feature



Fig 59

so-called idiopathic megaureters are in reality the only remaining signs of this type of obstruction In the majority of instances the cause of the obstruction remains a matter of speculation

We believe that such transient obstructions may occur from gumming up of the external meatus or even of the urethral lumen by embryonic adhesions comparable to the labial fusion so commonly seen in little girls or the preputio-glandal adhesions of the boy Such a condition operating *in utero* or in the neonatal period could exercise serious back pressure effects, and leave no other recognizable trace of its occurrence Any abnormal narrowing of the external meatus particularly as seen in hypospadias would predispose to temporary occlusion of this type. We have recently seen a striking example of such obliterative adhesions in the urethra at the age of 3 months

L.M. Female.

This baby presented as a case of acute retention and it was impossible to pass a catheter as no urethral orifice could be found Supra pubic cystotomy was therefore performed. In the acute stage of her illness she developed a large tender swelling in the right perinephric area which subsided on treatment with penicillin (*see* Fig. 58)

By the kindness of our colleagues in Cambridge, when the baby was sufficiently recovered she was brought up to Great Ormond Street. Under anaesthesia, and after some difficulty the orifice of the urethra was found in an ectopic position on the anterior vaginal wall, glued up by adhesions Eventually however a ureteric catheter was successfully passed This was retained for 5 days and on its withdrawal normal micturition was resumed. Intermittent dilatations have since been required

Birth injury does not seem to us to be an important factor in relation to urinary obstruction, though Spence (1933) has mentioned cases of spinal cord injury causing paralysis of the bladder together with paralysis of the skeletal muscles

In older children there is always the possibility that the passage of instruments may have ruptured unnoticed some mucosal folds, so that the retention is relieved before the cause can be found There is also the possibility that spasm of the internal meatus having for a time constituted an obstruction may subsequently relax. In this connection it is interesting to recall that infantile hypertrophic pyloric stenosis can undergo spontaneous cure, with resumption of the normal appearance of the pylorus (*see* Fig. 59)

In some cases there is a suggestion that redundancy of oedematous mucosa may be responsible for blocking the outlet of the bladder as in the following case

B G Male. Aged 4 months

This infant was admitted under Dr Evans at the age of 4 months on account of a discharge from the navel While in hospital he suddenly became very ill and had a high temperature, a distended bladder and 2 palpable kidneys The urine became infected and he was treated for a time with an indwelling catheter Normal micturition was, however not re-established on removing the catheter and the bladder was opened supra-pubically The vesical mucosa was congested and redundant and there was no evidence of a Marion type of obstruction at the bladder neck or in the posterior urethra.

A wedge was removed from the posterior lip of the internal meatus and the bladder closed with a supra pubic tube. The wound closed with some difficulty and the urine remained infected, but on discharge 6 weeks after the operation he was gaining weight

plication of the bulb with unabsorbable sutures, as suggested by Lewis (1949), has been undertaken with success A No 12 French rubber catheter is passed, and with the boy in the lithotomy position a mid-line perineal incision is made This is carried down as far as the bulb of the urethra, dividing the bulbo-spongiosus muscle which is retracted laterally Mattress sutures of No 3 Chinese silk are then inserted into the tissue of the bulb so as to plicate it tightly upon the catheter. These sutures may with advantage be placed in the tissue covering the ischio-cavernosus rather than in the spongy tissue The bulbo-spongiosus is then sutured with cat-gut and the wound closed without drainage The catheter can be removed at the end of the operation

THE NEUROGENIC BLADDER

Vesical retention of neurogenic origin requires special consideration The usual manifestation is incontinence with retention In rare instances, where the bladder neck is flaccid, there is continuous dribbling without retention

Distinction must be made from incontinence due to cerebral lesions associated with defective micturition control, for example cases of spastic paralysis (Little's disease) with mental retardation, and from functional micturition disorders (enuresis)

In the child the neurogenic bladder is almost always associated with congenital deformities of the spine, though acquired lesions such as transverse myelitis or compression paraplegia are sometimes encountered

Spinal defect

The spinal anomaly is most often some variety of spina bifida, and the site almost invariably lumbo-sacral or sacral Defects at higher levels are seldom attended by disturbances of micturition, or indeed by other neurological complications

Spina bifida may be accompanied by a meningocele or a meningo-myelocele, or may be only marked on the surface by a dimple, lipomatous mass or tuft of hair (spina bifida occulta) Lumbo-sacral meningoceles are not necessarily in the mid-line, and may present as eccentric cystic swellings, possibly filling out one or other buttock (*see* Figs 60 and 61) The true nature of the swelling may not then be appreciated

Gross congenital anomalies of the bony spine alone may also be responsible for nerve involvement affecting the bladder, absence of sacral vertebrae, hemi-vertebrae, for instance, have been found, and the absence of superficial evidence may be misleading (*see* Fig 62)

Neurological signs

The diagnosis of neurogenic bladder in the absence of other neurological signs should only be made with the greatest hesitation, the bladder does indeed show some characteristic and diagnostic features but a careful examination of the nervous system will almost always reveal some other evidence of the central nervous lesion A purely radiological spina bifida is a very common finding in children, and, even when associated with enuresis or with retention, should not be regarded as the responsible factor without corroborative evidence There is nothing to justify any lingering belief that these minor anomalies of spinal fusion have any causal relation to indeterminate disorders of micturition (enuresis)



FIG 60.—Neurogenic bladder Eccentric meningocele in left buttock Girl aged 1 year 2 months. Circum-anal anaesthesia and extreme degree of retention and upper tract involvement.

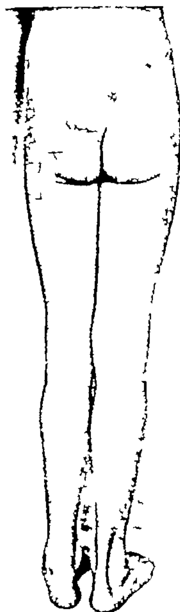


FIG 61.—Neurogenic bladder Eccentric meningocele (left sacral) Girl aged 11 years.

Two years before the girl in Fig 61 had been admitted to us removal had been attempted elsewhere. Since this operation she had suffered from urinary incontinence by day and night. On examination she was found to have saddle anaesthesia as indicated in the photograph, left pes cavus and a patch of anaesthesia on the lateral aspect of the left ankle. Residual urine 6 ounces. Bladder trabeculated, ureteric orifices normal. The cystogram showed no reflux. Training in manual expression has greatly improved her condition though the incontinence is not yet fully controlled.

Lesions of the sacral nerve roots are naturally the commonest associations of urinary symptoms, for example a saddle area of anaesthesia and relaxation of the anal sphincter Only on very rare occasions does the anus escape when the urinary sphincters are affected or *vice versa* Pes cavus and patches of anaesthesia around the heel are common, and in the severe cases there may be complete paraplegia



FIG 62 —Neurogenic bladder Straight skiagram showing congenital lumbo-sacral bone defect in a girl aged 4 years without any associated external swelling

This child had been noticed to have continual dribbling incontinence of urine since birth and only a very small amount was passed at micturition The bladder was distended and manual expression of urine easy The anal sphincter appeared normal but she suffered from obstinate constipation There was a left-sided congenital dislocation of the hip, absent left knee and ankle jerks and paresis of all the left lower limb muscles Intravenous pyelogram showed gross upper tract dilatation The residual urine varied considerably from time to time, and she suffered several attacks of uraemia with severe pyocyaneus infection, each time being relieved by a period of catheter drainage and bladder wash-outs Finally a bladder-neck resection was performed with some improvement

In spina bifida occulta it is well known that the neurological signs may first appear in later childhood, a fact attributed to the increasing tension produced at the site of the defect by the disproportionate growth of vertebral column and spinal cord

It may be remarked that independent deformities, for example talipes, of non-paralytic origin may be associated with spina bifida

Urological features

In the severe lesions dribbling incontinence of urine and the absence of any definite act of micturition are noted from the time of birth The bladder can usually be felt to be distended and firm, urine is easily expressed from it by manual pressure, and ammoniacal burns and even ulcerations are evident around the genitalia In the less severe cases some urine can be passed as a stream, but incontinence both by day and night is a troublesome feature

In many cases, especially those without any paralytic signs in the legs, the urinary symptoms are not noticed until the child is past the infant years The symptoms in infancy may of course have escaped notice, but this is not necessarily so There can be no doubt that in some instances the condition does actually

deteriorate A child who has had no more than frequency and occasional accidents may fall back to continual dribbling incontinence by day and night. In some cases this deterioration may be traced to the onset of infection but in others the cause is obscure and the possibility of progressive nerve damage cannot be excluded Ill advised neuro-surgical operations unquestionably worsen the condition

Acute retention is rare but may be seen after operations upon the spine, and has been observed upon one occasion in a boy with severe constipation The accumulation of excessive amounts of faeces in the rectum always exacerbates the symptoms in mild cases

Some residual urine is present in almost every instance but it varies considerably in amount from case to case The bladder is always trabeculated and often sacculated on the cystogram it may appear as a large lax organ with a "cottage loaf" outline or may be comparatively small and tapering towards the apex ("fir-cone" appearance) (*see* Fig. 63) Vesico-ureteral reflux is a marked feature occurring earlier and more commonly than in simple obstructions it often affords



FIG 63.—Neurogenic bladder Cystogram from girl aged 11 years.

Sacral meningocele operated upon at 3 months. Since that time had been suffering from continual dribbling of urine and obstinate constipation. No improvement had been obtained by urethral dilatation, anti-spasmodic drugs or bilateral sympathectomy The urine was infected with *B. coli* Blood urea was 40 mg. per 100 ml. There was sensory loss in the area supplied by S.3 and 4. Cystoscopy showed gross trabeculation of the bladder which contained 18½ ounces residual urine, but there was very little vesico-ureteral reflux. Intravenous pyelogram showed normal right kidney but only a few dilated calyces were outlined on the left. A wedge resection of the bladder neck was performed and manual expression practised. The residual urine fell quite rapidly to 1 ounce. She could be kept almost dry by day and night when seen 18 months later

a very complete depiction of the dilated upper tract on cystography (*see* Fig. 65) Relaxation of the bladder neck with filling of the dilated proximal urethra is seen in many cystograms both in girls and boys, even in those cases in whom the internal sphincter appears as a prominent ridge during cystoscopy (*see* Fig. 64) Sensation of bladder fulness is for the most part absent, though the milder cases may have some feeling of distension and even of urgency

Attempts are sometimes made upon the basis of symptoms and of cystometrograms to locate the nerve lesion in the anterior or posterior roots, and to distinguish the irritative from the paralytic effects Thus in the large atonic type of bladder, having a general similarity to the typical tabetic organ, posterior root damage is thought to predominate, in contrast to the small high-tension bladder, which simulates that seen after traumatic section of the cauda equina (Langworthy, 1936,



FIG 64 —Neurogenic bladder
Girl aged 4 years

Under treatment by Mr Eric Lloyd for multiple skeletal deformities and paralysis *Spina bifida occulta* deformity in the dorso-lumbar region and the scar of previous operations in the cervical region Urinary symptoms were frequency with mild occasional incontinence by day nocturnal incontinence usually but not invariably Bladder not palpable A cystogram showed reflux into dilated left ureter and kidney and filling of the upper urethra Controlled by training



FIG 65 —Neurogenic bladder
Girl aged 5 years

Sacral meningocele excised at 1 year Continual incontinence, bladder and right kidney palpable, residual urine 9 ounces Trabeculated bladder with reflux into dilated ureters Regime of manual expression no improvement Bilateral lumbar ganglionectomy very little change One year later residual urine 10 ounces After bladder neck resection this child's condition improved Residual urine reduced to ½ ounce

Riches, 1944) As with other types of neurogenic bladder, however, such distinctions can seldom be clear-cut and are not of great assistance in treatment

Infection is always present in long-standing cases and the urine may be heavy with ropy mucopus Infection causes a deterioration of bladder function, an increase in upper tract dilatation and progressive damage to the renal parenchyma, which is frequently the cause of death in these unfortunate children

Diagnosis

Diagnosis rarely presents any difficulty except in the mild cases The presence of the spinal deformity together with the accompanying signs in the central nervous system are in themselves conclusive enough In any doubtful case, however the type of bladder is in itself characteristic In no other type of retention is manual expression of the urine so easy vesico-ureteral reflux so early and so striking, or trabeculation and the bar at the bladder neck so prominent a feature

Treatment

Direct neurosurgical operations upon the meningeal deformity or upon the nerve roots have been in the cases known to us, uniformly disappointing both in the prophylaxis and treatment of urinary symptoms In the new born infant operation for meningocele may be required but the decision is no easy one to make and the indications for it are not urological Where there is gross paraplegia and flaccid sphincters the prognosis is obviously quite hopeless and operation unjustifiable but in many cases the prospect seems more favourable the limb movements are good and the rectum and bladder apparently controlled When the sac is large thin walled and tense, so that rupture seems imminent, there is a strong temptation to undertake immediate operation to save the baby s life Unfortunately the risks of such an operation are by no means inconsiderable Apart from the danger of development of hydrocephalus the interference may well increase the nerve damage and precipitate urinary symptoms. Moreover if left to themselves these thin walled sacs show a surprising tendency to shrivel and their coverings to thicken up It is true that many of these cases even if not operated on ultimately develop urinary symptoms, but we must record our conviction that operation is only too likely to be detrimental to bladder function

In spina bifida occulta operations are sometimes advised to free the nerve roots We have not seen good results from this practice and Elsberg (1941) states that in long-standing urinary disorders he has not obtained any significant improvement after such operations

Treatment directed towards the management of the bladder is of considerably greater value The object of such treatment is to control the infection and to reduce to a minimum the residual urine Although it is unlikely that complete continence will be obtained the leakage can often be brought within manageable proportions by keeping the volume of urine in the bladder very small Control of the infection is carried out with the aid of chemotherapy but in neglected cases a course of bladder lavage with appropriate antiseptic lotions is an essential preliminary The child with the help of the mother is then taught to empty the bladder at regular intervals by manual expression and the residual urine measured after this procedure At the same time a regular bowel action must be obtained with the assistance, if need be of rectal wash-outs If after 1–2 weeks on this regime a considerable amount of residual urine is still found a wedge resection of the bladder neck may be performed This operation even in cases where dilatation of the proximal urethra has been noted in the cystogram has the effect of reducing the residual urine after manual expression with a corresponding improvement in the incontinence Its effect is perhaps greater where the residual has been very large than in the small high tension bladders

We have not found sympathectomy to be of any value in restoring bladder function, and, in the absence of a normally acting detrusor muscle, operations for incontinence which involve increase of the urethral resistance (for example plication of the bulb) are very seldom effective

Where all other measures have failed, some form of portable urinal must be ordered, but in girls at least such apparatus is seldom satisfactory

Clearly the full co-operation of the child is essential to the successful management of the neurogenic bladder, and it is unfortunate that the child's disability has too often led to isolationism; and the psychological barriers which have been erected are difficult to overcome

CHAPTER 9

URINARY OBSTRUCTION—THE DILATED URETER

INTRODUCTORY CONSIDERATIONS

Obstruction at the uretero-vesical level

We now come to consider the group of cases in which the site of obstruction is at the uretero-vesical level and in which the ureter is the outstanding concern This is a very large and heterogeneous group which includes a number of developmental anomalies, and poses many of the most difficult problems in diagnosis and treatment in the whole field of children s urology It must be admitted that the obstruction element is often elusive In the main however it is the hold up of the urinary flow which determines the deleterious effects of the great majority of these lesions and it is therefore convenient to retain the term obstruction though it may not be universally accurate

The outstanding feature is dilatation

Factors affecting ureteric calibre

Tone

That the ureter is capable of rapid and striking changes in size and form is a matter of common observation particularly during operations on children In normal circumstances, the ureter is clearly capable of considerable dilatation and may appear quite large in relaxation contracting down to a much narrower calibre in the systolic phase These variations in calibre are brisk and appear distinctive in type, and may conveniently be considered as variations in tone.

Propulsion

The propulsion wave is of a different nature and is comparab e to cardiac con traction We were afforded a remarkable demonstration of this propulsive action during life in the case depicted in Fig. 20 (*a*) and (*b*) page 79 In the course of the operation on this child the agenetic kidney and upper ureter were revealed Approximately two-thirds of the lower ureter appeared normal as shown On accidental forceps stimulation of the upper end of this normal segment the ureter shot down into the pelvis out of sight by a striking piston like action, returning on relaxation The same response was repeated each time the stimulus was applied No doubt the absence of any normal fixation above permitted undue exaggeration of the excursion but the demonstration of the piston like propulsion wave was instructive This clearly is the type of contraction which propels the efflux as seen in cystoscopy

All neuro-muscular activities in the young are relatively excitable and brisk and the ureter is no exception We imagine in consequence a certain inherent

susceptibility to peristaltic disorders (spasm and so forth) in infancy and childhood, which, coupled with the narrow calibre of the tube and the readiness with which it may be "gummed up" by inflammatory reaction, constitutes a factor of great importance in relation to persisting infections and their after effects



FIG 66—Megaureter (probably *Béance congenitale*) Reflux pyelogram from an infant girl aged 1 year Acquired obstruction at uretero-pelvic junction

This infant had been under the care of Dr Lightwood for recurring attacks of severe urinary infection On admission the urine was heavily infected Cystoscopy showed a bullous cystitis and both ureteric orifices large and patulous with no efflux from the left Retrograde pyelography showed gross dilatation of both ureters, the right pelvis normal but the left did not fill The radiography indicated a persisting block at the left pelvi-ureteric junction and it was thought that the hold-up at this point was responsible for the persistent infection The left kidney was explored and inflammatory adhesions involving the PU junction were freed Following this operation the infection cleared In the subsequent 6 years the child has progressed normally and has had no further symptoms She is in fact so well that her parents will not allow re-examination of the urinary tract, and the classification of this particular megaureter remains in doubt

Paralytic ureterectasis

An important result of infection on the ureteral wall is, in our view, that it induces a paralysis of the tone element, leading to a condition of paralytic atony and distension analogous to that seen in the small intestine in paralytic ileus though more chronic in nature Particularly is this seen in the lumbar segment (*see* later)

Evolution and pathology of ureteral dilatation

The effects of increased intraureteric tension are like those seen in a vein becoming varicose an initial hypertrophy in the effort to overcome the resistance to normal emptying, followed by a breakdown expressed in dilatation and thinning of the wall increase in capacity by lengthening and growing tortuosity Infection and periureteritis ultimately fix the ureter and glue the pouches to surrounding tissues and to one another so that they become inert pockets of stagnant purulent urine Any sort of natural drainage of such a ureter becomes a physical im possibility

The secondary effects on the kidney follow from back pressure and spread of the infection The pelvi ureteric sphincter does seem to protect the kidney from back pressure effects with considerable efficiency but severe infection must exercise a most sinister influence

Adhesions around the pelvi ureteric junction may occasion a secondary obstructive hydro-nephrosis and this may very rapidly intensify the damage to the kidney (*see* Fig 66)

URETERECTASIS

Ureteral dilatation of moderate degree is seen in some phases (developing or otherwise) of megaureter of whatever nature Under the heading of Ureterectasis, however are included certain instances of moderate dilatation particularly of the lumbar segment, which are frequently seen in practice and are of a different type. The lumbar segment appears to be very susceptible to atony This is notably seen in the hormonal atony of pregnancy Infection determines a similar effect Lumbar ureterectasis often unilateral is observed in children commonly girls with recurrent infection it is seen below septic calculi in the renal pelvis and below infected hydro-nephrosis A study of our cases shows the dilatation to be variable, and in favourable circumstances, recoverable Thus in serial skiagrams it is characteristic to see at times a ureter of normal calibre, at others a well marked dilatation

The dilatation may disappear promptly after removal of the source of irritation and infection, for example a calculus, and sterilization of the urine (*see* Fig. 67 (*a*) and (*b*)) or it may persist for a year or 18 months or even longer Given control of the infection however there is a tendency to recover

As already stated we incline to regard the dilatation in these cases as an expression of ureteral ileus of inflammatory origin rather than the result of spasm which could hardly be so consistently localized at the pelvic brim

In hydro-nephrosis it is noteworthy that the infected cases tend to be com paratively painless, which suggests that the infection exercises a similar weakening or atonizing effect on the muscle of the renal pelvis

Disuse atony

Lumbar ureterectasis is also seen in some instances apart from infection notably below a sterile hydro-nephrosis or aplastic kidney Here the dilatation appears to be due to disuse atony At operation contraction appears normally brisk on stimulation and the condition seems to be simply explained by absence of the normal stimulation of the pelvic efflux

3 Megaureter due to recognizable organic obstruction of the ureter itself
4 Megaureter with apparently normal uretero-vesical junction and lower urinary tract
5 Megaureter—bilateral with reflux and ear trumpet orifices —*Béance congenitale* (Marion 1940)
6 Megaureter associated with duplication

1 Megaureter resulting from back pressure due to obstruction at the bladder neck or below

In cases where vesical retention is evident, the secondary nature of the ureteric dilatation will not be in doubt in certain circumstances however megaureter may be the predominant feature of the case and the primary vesical disorder is apt to be overlooked It has already been mentioned (Chapter 8) that in the child detrusor hypertrophy may be sufficient to overcome the obstruction and empty the bladder although the upper urinary tract shows signs of back pressure In such a case intravenous pyelograms reveal dilatation of one or both ureters and a hold up where the hypertrophied bladder muscle compresses the intramural ureter The cystogram then seldom shows a reflux and the ureteric orifice is not dilated

Attention has also been drawn to the transient nature of some infra vesical obstructions in infancy it is suggested that in these cases normal micturition may be re-established but that ureteric dilatation may remain The history will perhaps betray the nature of the primary lesion, or the cystoscopic picture of dilated railway tunnel orifices may be suggestive

The distinctive features of this type of megaureter are therefore as follows

(a) *Clinical*—Slight difficulty in micturition a poor stream or a palpably thickened bladder with a few ounces of residual urine may be noted (*see* Fig 48) Alternatively micturition may be entirely normal at the time of examination, but a transient attack of retention is mentioned in the history (*see* Fig. 68)

(b) *Cystoscopic*—In most cases trabeculation is a marked feature In the rarer instances where the obstruction has been transient or has been relieved trabeculation may be slight or even absent. The ureteric orifices may appear normal but more usually are distinctly large slits and in many cases are of the railway tunnel type that is a large black hole retaining the normal obliquity and outline of the ureteric orifice (Fig. 69)

Vesical diverticula are exceedingly rare in children but a true diverticulum if seen affords decisive evidence of lower urinary obstruction even in the absence of residual The true diverticulum which has drawn the ureter into itself must be distinguished from the ear trumpet orifice (*see* later) In the former case the interureteric bar can often be followed over the edge of the diverticulum and the orifice lies just inside on the medial wall of the sac

(c) *Radiological*—The cystogram commonly shows reflux of varying degree though as already stated this feature may be absent The ureterogram shows the dilatation to extend completely down to the bladder without any relative narrowing of the lower end, except possibly the intramural portion The dilatation is always bilateral but one side may often be much more advanced than the other a well known feature of the back pressure ureter resulting from any lower urinary obstruction. The degree depends upon the circumstances of the individual cases

with, of course, corresponding variations in the degree of involvement of the renal pelvis and calyces

These cystoscopic and radiological appearances are most typically seen in the gross megaureters of prolonged vesical retention (vesical neck and post-urethral obstructions and in the neurogenic bladder)

It is obviously important to distinguish this type of megaureter from one in which the primary disorder is at the uretero-vesical junction (*see* later) Careful

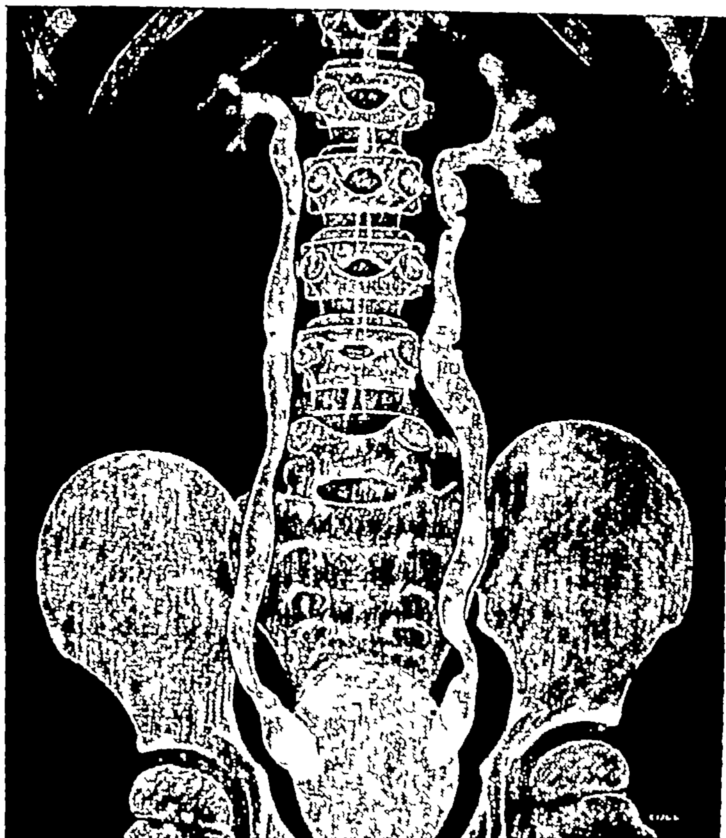


FIG 68 —Megaureter Back pressure from vesical retention (Type 1) Pyelographic tracing from a boy aged 7 years

First admitted under Dr Lightwood with a diagnosis of encephalitis The child was extremely ill for some days and unconscious for a part of the time During this period he had retention necessitating catheterization After 2 months he was discharged with no symptoms except a residual squint One month later, however, he had an attack of abdominal pain with fever and cloudy urine Again he had retention requiring catheterization When at a later date urological investigation was undertaken he had normal micturition and no residual urine Intravenous pyelogram showed considerable dilatation of both ureters with poor function of the right kidney Cystoscopy showed no trabeculation, a normal mucosa and no change at the bladder neck Both ureteric orifices were widely dilated but retained their full obliquity ("railway tunnel") The orifice contracted with the efflux The child has been symptom free for the past 18 months, the urine is sterile and the ureteric dilatation remains unchanged

attention to the cystoscopic and radiological appearances outlined above usually enables this distinction to be made

Treatment and prognosis

In cases with obvious vesical obstruction the treatment is that of the particular cause.

In bilateral cases even where there is uncertainty as to the presence of vesical neck obstruction it may be thought justifiable to open the bladder and excise a wedge of the neck

Where there is a history but no remaining sign of obstruction we may assume that the megaureters are stabilized and that no further dilatation will occur These should be treated conservatively and the prognosis is good in the absence of serious renal damage

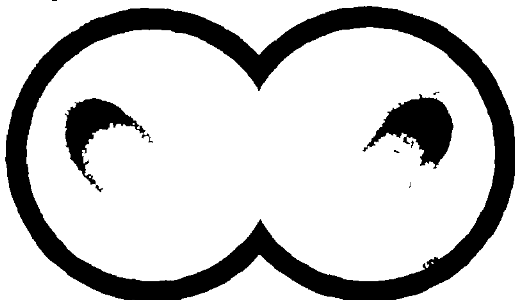


FIG 69 —Drawing to illustrate cystoscopic appearance of the railway tunnel type of ureteric orifice.

2. Megaureter resulting from cystitis

There is a comparatively large group of cases chiefly girls in whom a moderate degree of bilateral megaureter is found to complicate a lasting or recurrent cystitis In most a vesico-ureteral reflux has been noticed when the disorder was first diagnosed, but later in the course the reflux is no longer present There can be no doubt that severe oedema around the ureteric orifice can render it so rigid as to disorganize the normal uretero-vesical mechanism and determine reflux This, together with the ileus effect of inflammation probably accounts for the dilatation in this group

Megaureters of this type are normally bilateral but one may be more advanced than the other The dilatation remains moderate in degree and if the infection subsides becomes stationary and may even show some tendency to regress

The distinctive features are

(a) *Clinical*—Severe lasting or recurring cystitis absence of any signs of lower urinary obstruction

(b) *Cystoscopic*—The bladder shows cystitis in varying degrees In the chronic stage cystitis cystica has been observed The orifices at first may appear oedematous and may be pouting. Later with subsidence of the infection they may return to a normal appearance even though the ureter above is still dilated. Sometimes they have been noted to appear normal at rest but to open widely and slowly at the time of the efflux assuming the appearance of a large black hole (*see* Type 5)

(c) *Radiological*—The dilatation is moderate, extends through the length of the ureter down to the bladder (*see* Fig 70) Kinks are not a very marked feature but at the pelvi-ureteric junction they may become fixed by the inflammation and be responsible for a retentive hydro-nephrosis which may maintain the infection

It may be emphasized that the bladder in these cases of Type 2 megaureter retains a reasonable capacity Ureteric dilatation of considerable degree also accompanies the severely contracted bladder from whatever cause Thus in unilateral renal tuberculosis dilatation of the contra-lateral renal pelvis and ureter may be seen as a result of the systolic bladder, and the same effect can be observed in long-standing cases of vesical calculus In such cases bladder capacity is so small as to be distinctive



FIG 70—Megaureter (Type 2) Excretion pyelogram from a girl aged 9 years Bilateral dilatation, most marked on right side

Investigated on account of enuresis and urinary infection Cystoscopy showed a basal cystitis but normal UOs The urinary infection was controlled after some time and 3 years later there were no urinary symptoms An intravenous pyelogram showed the ureteric dilatation unchanged

Treatment and prognosis

The essential objective is to sterilize the urine and keep it so If sterility can be maintained the prognosis is good, even though considerable dilatation persists Secondary effects (in particular fixation adhesions at the pelvi-ureteric junction with hydro-nephrosis) may require operative interference

3 Megaureter due to recognizable organic obstruction at ureteric orifice (stricture, ureterocele)

This is a somewhat heterogeneous group, but the cases have in common an obstruction at the uretero-vesical junction which can be recognized cystoscopically. Impacted calculus must also be considered in the differential diagnosis, though the resulting ureteric dilatation will normally be more acute than in the common megaureters

Stricture

This may be congenital or acquired

(a) *Clinical* —Pain is more likely to be a prominent feature in this group though not necessarily so It will be colicky in nature vaguely abdominal in character notoriously difficult to evaluate in childhood and misleading to the unwary Chronic or recurring infection is a very likely presenting symptom An impacted stone in the lower end of the ureter may be felt per rectum

(b) *Cystoscopic* —Stricture of the orifice is by no means easy to be sure of Many ureteric orifices in childhood look deceptively small The appearances vary a good deal with the degree of distension of the bladder Inability to pass a small ureteric catheter is not a reliable guide. an orifice which looks small may be found easily negotiable, while one which looks normal may not. A true pin hole meatus may be detectable only by spotting the efflux and in our experience it declares itself most often as a ureterocele An acquired stricture is more easily recognized in the rare instances we have seen by the scarring and rigidity The presence of an impacted stone may be recognized by the usual angry oedematous bulge of the ureteric orifice.

(c) *Radiological* —The dilatation is typically fusiform extending at its apex to the site of the obstruction

Treatment and prognosis

The clear indication is to relieve the obstruction Dilatation of the narrowed orifice may be attempted cystoscopically By this means the orifice may be incised by diathermy and dilated by graduated bougies but it should be recognized that such procedures have very limited possibilities in childhood and we have not been impressed by their efficacy

A satisfactory ureteric meatotomy is best achieved by open cystotomy permitting of adequate surgery under direct vision Genuine strictures are not too easily relieved and subsequent recontracture is very apt to occur The simplest procedure is free incision by diathermy in an upward and outward direction followed by full dilatation with graduated bougies This may not be enough and total excision of the orifice and intramural ureter with re implantation of the pull through type may well come to be more extensively practised in future

The removal of impacted stones must always be followed by adequate dilatation and it is no easy matter to ensure this and prevent recurrence. Subsequent nephro-ureterectomy has been necessary in several of our cases.

Provided the obstruction be adequately relieved and the infection controlled prognosis is not unfavourable

Ureterocele

Ureterocele may suitably be included in this section since there is evident pathology at the lower end of the ureter and it seems likely that a congenitally stenotic pin hole orifice is often responsible for the lesion The essential feature of a ureterocele is cystic dilatation of the intravesical portion of the ureter which balloons into the lumen of the bladder carrying up on its anterior surface the stenosed ureteric orifice The cyst is lined by ureteric epithelium and covered by vesical epithelium its wall varies somewhat in thickness but almost always contains some muscular tissue presumably derived from those longitudinal fibres which accompany the intramural ureter as far as its termination Proximal to the cystic dilatation the ureter is narrowed as it passes through the muscular coats of the

bladder, but the obstructive effect of the lesion is evidenced by a variable degree of dilatation of the extravesical course of the ureter As was first noted by Hellstrom (1937), the pelvic segment is always the first to show definite dilatation, a feature also of the Type 4 megaureter (*see* page 146) At a later stage the upper ureter and pelvis may also be involved and the urinary stasis may well be complicated by the formation of stones

The ureterocele may attain a considerable size and by impinging upon the internal meatus produce a mild degree of vesical obstruction Many cases have been reported in girls in which prolapse through the urethra has occurred and sloughing of the extruded cyst wall may then follow Bilateral ureterocele is not uncommon.

(a) *Clinical*—The symptoms of ureterocele are in no way characteristic Pain on the affected side, frequency, enuresis and occasionally urinary infection may be noted

(b) *Cystoscopic*—At cystoscopy the moderate sized translucent cysts are recognized without difficulty, situated at the site of the ureteric orifice and distending with each contraction of the ureter The efflux from the stenotic orifice may be seen as the cyst deflates In early cases the orifice appears flat and normal when at rest and balloons only immediately before the efflux Very large ureteroceles are sometimes difficult to diagnose because it is impossible to get the cystoscope sufficiently far away from them to recognize their true nature The translucency of a ureterocele is often deceptive, and it may be found at operation that the wall is considerably thicker than was supposed at cystoscopy. Ureterocele must be distinguished from the so-called prolapse of the ureter in which the orifice appears to be situated upon the summit of a pyramidal mound but in which there is no suggestion of cyst formation This appearance may be seen in the absence of any other pathology but has been observed by us on one occasion in association with a Type 4 megaureter

(c) *Radiological*—Provided renal function is unimpaired, the intravenous urogram gives a diagnostic picture A circular area of increased density is seen within the bladder shadow and is surrounded by a halo of comparative translucency The dilatation of the pelvic ureter may be seen, and where the whole course of the lower ureter is outlined it has a characteristic "cobra head" appearance (*see* Fig 71).

The true ureterocele, which has a comparatively narrow base, must be distinguished from the broad intravesical protrusion of the lower end of an ectopic ureter Mertz, Hendricks and Garrett (1948) have described a variety of ureterocele, upon which no ureteric orifice could be found, under the name "cystic uretero-vesical protrusion". In our experience a ureteric orifice is present in such cases, although it may be in the urethra or even further afield and may be stenotic; the cystic protrusion is simply the bulging of the dilated lower end of the ureter which lies in close proximity to the bladder mucosa Indeed it is difficult to believe that the kidney would continue to secrete urine were no outlet present This type of cystic protrusion does not have the characteristic translucency of the true ureterocele, and its broad base is often prolonged down towards the bladder neck, which it may deform As described in Chapter 7 the ectopic ureter is most often seen in girls and is commonly derived from the upper pelvis of a double

kidney (deuteropyelon) Ureterocele may occur in cases of complete pyelon duplex, where both ureters open into the bladder but again the orifice of the deuteropyelon is always the one involved and the cyst is apt to be more broad based than the classical ureterocele In the latter form the orifice of the protopyelon is often dragged up on to the slope of the cyst and may be dragged open so that it allows free vesico-ureteral reflux. The obstruction to the bladder neck may result in some dilatation of the contra lateral ureter



FIG 71 —Megaureter with ureterocele (right) Type 3 Excretion pyelogram from a girl aged 2 years

Investigated for recurrent pyuria. Cystoscopy confirmed the diagnosis The ureterocele was excised at cystotomy and when seen 6 months later the child had had no further symptoms.

With regard to the pathogenesis of the true ureterocele in which ureteric ectopia is not a factor it would seem that a congenitally stenotic orifice is the usual cause Acquired stenosis such as may follow the passage of a stone does not have the same effect because the accompanying fibrosis tethers the orifice to the underlying bladder musculature Dilatation then only involves the extravesical ureter The closing membrane first described by Chwalla (1927a) which occludes the uretero-vesical junction in early foetal life (*see* page 65) does not appear to be connected with the formation of a true ureterocele which does not in any way suggest a bulging membrane and upon which a ureteric orifice can always be found.

Treatment

The obstructive factor may be removed by excision of the cyst but as in all types of megaureter which have their origin in early childhood complete regression of the ureteric dilatation is not to be expected *Transurethral* methods of destroying the cyst are seldom applicable to small children as a simple puncture with the

diathermy electrode is insufficient and the instruments required for more radical procedures are too large At open operation through the bladder it is a simple matter to cut away the redundant mucosa, and if necessary control the bleeding with a running suture of unhardened cat-gut Where there has been severe *infection nephro-ureterectomy, or in the case of double ureters hemi-nephrectomy,* may be required

4 Megaureter with apparently normal uretero-vesical junction and lower urinary tract

In this group we would include a type of megaureter which appears to us distinctive in that typically it is unilateral and confined to older children, usually boys Admittedly it has some resemblance to those with stricture (Type 3) and, if the bilateral cases are placed in this group, such instances are very difficult to distinguish from some types of lower urinary tract obstruction with intact ureteric orifices in which the megaureter is attributed to vesical hypertrophy (Type 1)

In the cases now under consideration there is no evidence of such hypertrophy of the bladder, the ureteric orifice appears perfectly normal The dysfunction is at present inexplicable and there is no alternative but to call it an achalasia

(a) *Clinical*—The presenting feature is frequently pyuria, or recurrent fever and aching pain in the side The sharp attacks of pain and vomiting, seen in simple hydro-nephrosis, do not occur In other cases the urine is sterile Two such children, while in hospital for an unrelated complaint, each had a single short attack of haematuria. On investigation each was found to have a unilateral megaureter of this type, with no detectable abnormality at the lower end, and no evidence of stone One has persisted unchanged for 3 years The other, in whom attempts were made to dilate the orifice, became infected and was later treated by nephro-ureterectomy (*see* Fig. 72)

(b) *Cystoscopic.*—The bladder and ureteric orifices appear perfectly normal and catheters pass without difficulty

(c) *Radiological*—The dilatation ends at a point above the bladder so that the terminal segment of the ureter, usually up to an inch or so in length, is of normal calibre The narrow portion may be shorter and sometimes appears confined to the intramural ureter

Treatment

When the urine is sterile, deterioration of renal function is very slow, and in mild cases it may be wise to refrain from any procedure likely to introduce infection No specific treatment is then given but the progress of the condition watched by periodic intravenous pyelograms

Where the dilatation is considerable but the kidney good and the urine sterile or sterilizable, conservative surgery is indicated division of the ureter and re-implantation, or possibly excision of the ureteric orifice, and a "pull through" procedure A simple lateral anastomosis between the dilated segment of the ureter and the bladder is almost valueless since it leaves the dilated pouch at the lower end undrained, and infection is then difficult or impossible to eradicate Where re-implantation is decided upon the ureter should be cut across where it enters the bladder, or at least through the narrow segment The kinks should be

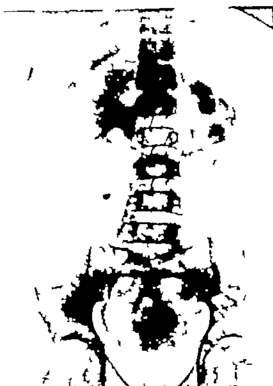


FIG. 72.—Megaureter (Type 4) Excretion pyelogram showing gross dilatation of the left ureter and kidney from a boy aged 12 years.

This child while recovering from an orchidopexy had an attack of haematuria. Cystoscopy showed normal bladder apart from slight oedema of the left ureteric orifice. Urine sterile. A transvesical ureteric meatotomy was performed but the symptoms recurred later and nephro-ureterectomy was ultimately necessary. Following this, no further symptoms.



FIG. 73.—Megaureter (Type 4). Specimen of right kidney and ureter from a boy aged 6 years.

Investigated for enuresis and an enlarged right kidney. Urine sterile. Intravenous pyelogram showed normal left kidney but no apparent function on the right side. At cystoscopy the bladder was normal and a retrograde pyelogram showed an enormously dilated right ureter. A nephro-ureterectomy was performed, which included the narrow terminal ureteral segment (q [illegible]). The child had no further symptoms (3 years).

straightened by division of adhesions and the ureter may then be found unnecessarily long in which case the lower end should be amputated, leaving sufficient length for a comfortable anastomosis without tension. Where the dilatation has been very great it is wise to excise a strip from the lateral aspect of the ureter in order to render the lower end fusiform, as advised by Crabtree (1937) Re-implantation of a dilated ureter is almost always followed by vesico-ureteral reflux, but this is not necessarily deleterious provided the urine can be sterilized

Where the dilatation is considerable, infection severe or renal damage serious, nephro-ureterectomy is the treatment of choice provided the disease is unilateral There is always a certain anxiety that the contra-lateral ureter will become dilated subsequent to the operation, but we have had no such disappointing experience hitherto, and it would seem that there is no need for undue apprehension (*see* Fig 73)

Discussion

Beer and Hyman (1930) were of the opinion that this type of case is due to the passage of a ureteric calculus, but the absence of any history of stone, the persistence of the dilatation in the absence of a stricture and the comparative rarity of small sterile stones in children render this theory improbable Most authors regard these cases as congenital, either in the sense that there is a congenital dysfunction at the uretero-vesical junction producing obstruction or that there is a congenital weakness, atony, of the ureteric muscle The evidence, though not conclusive, appears to favour the view that the narrow and apparently normal lower end constitutes in some way a functional obstruction The muscle layers of the dilated portion are always hypertrophied, though to a varying degree Sometimes with gross dilatation the wall appears thin, but upon deflation the hypertrophy will be evident and unless paralysed by infection or fibrosis even very large ureters retain a surprising contractility The fact that the dilatation often stops short of the renal pelvis does not, we believe, indicate any unusual deficiency in the ureter, since this limitation may be a feature of all types of mild ureteric obstruction which arise during early childhood

The analogy to the colon in Hirschsprung's disease is, of course, very striking In the latter Bodian and Stephens (1949) have stressed the absence of ganglion cells in the non-functioning recto-sigmoid segment distal to the dilatation, and their findings afford some rationale for the operation of recto-sigmoidectomy which is now in vogue and of which the immediate results are so gratifying Since in the type of megaureter now under consideration there is also a narrow segment at the lower end, it is tempting to suppose that an absence of ganglion cells might again be responsible However, ganglion cells are not normally to be found in the intramuscular layers of the ureter, so that a strictly analogous disease is not possible, and there is as yet no evidence for a ganglionic deficiency at any more distant point It is often stated that megaureter is commonly associated with the megacolon in Hirschsprung's disease, but we have not found this to be true In all our cases of Hirschsprung's disease explored and treated surgically, the ureters were visibly normal in calibre It is possible that in long-standing cases of megacolon, some dilatation of the ureters may occur from pressure of the colonic sac.

In our experience the treatment of this type of megaureter by drugs such as Pituitrin, Syntropan mecholyl bromide and by the administration of spinal anaesthetics has been valueless For these reasons we do not consider that the condition has a neurogenic basis

Vascular and vas deferens compression
Obstruction of the pelvic ureter by compression of the uterine vessels in girls or the vas deferens in boys is described (Campbell, 1937) We have not been able to recognize any instances of this but the possibility should be borne in mind

5 **Megaureter—bilateral with reflux and ear trumpet orifice**—*Béance congenitale*
This group includes a number of the most dramatic bilateral megaureters of child hood They come to notice most often in infancy and most of the instances we have seen have been in baby girls Some have been under observation for many

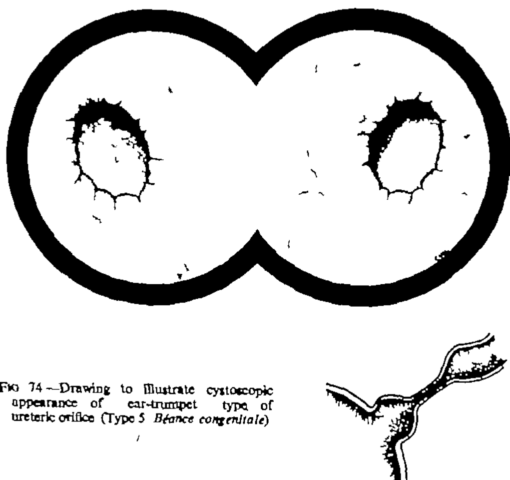


FIG 74—Drawing to illustrate cystoscopic appearance of ear-trumpet type of ureteric orifice (Type 5 *Béance congenitale*)

years All have fundamental characteristics in common The condition is con genital and bilateral with distinctive ureteric orifices remarkably free reflux and gross dilatation there is no trace of infra vesical obstruction Any minor variations there may be in individual cases can be explained by differences of degree age and the modifying effects of severe infection. The distinctive features are:

(a) *Clinical*—The story dates from early infancy and is one of severe infection persisting chronically and punctuated by acute "flares" which may be devastating Many of our cases have been seen in the first year of life In older children, seen then for the first time, even though we must assume a milder degree of the condition, some such long-standing history is usually forthcoming There is never at any time any evidence of vesical retention or of primary cystitis Cysto-urethrography shows no abnormality at the bladder neck or in the urethra

(b) *Cystoscopic.*—The vesical mucosa shows no abnormality unless there be a secondary cystitis, and there is no trabeculation The ureteric orifices present a remarkable appearance (*see* Fig 74) Each is represented by a large round hole,

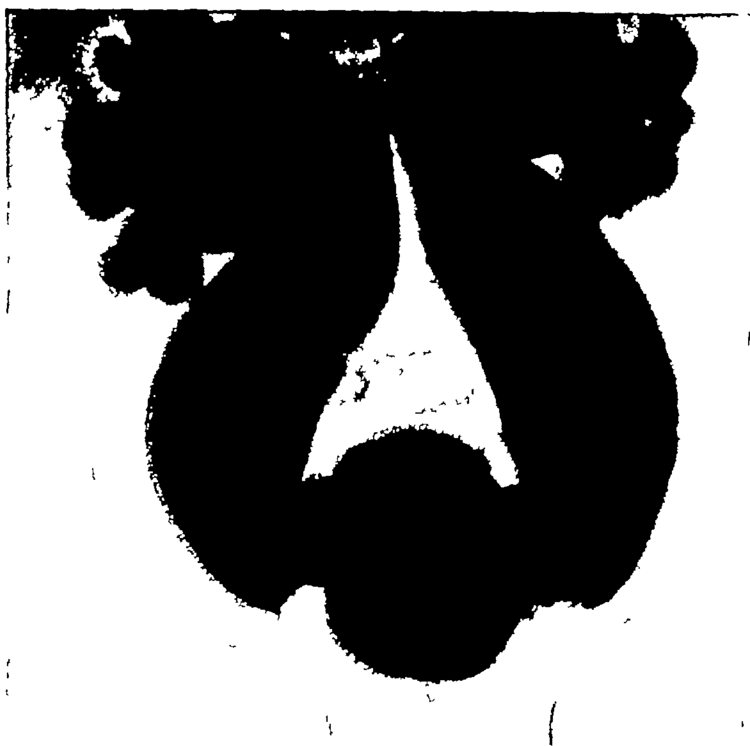


FIG 75—Megaureter (Type 5) *Beance congenitale* Voiding cysto-urethrogram from a baby girl aged 1 year 4 months

Referred by Dr Sheldon on account of recurrent urinary infection since the age of 8 months On examination urine sterile, micturition normal Blood urea 38 mg per 100 ml Cystoscopy mucosa normal, no trabeculation The ureteric orifices were represented by large round holes within which the ureteric lumen could be seen disappearing upwards ("ear-trumpet" type) There were no neurological signs, and a cystogram repeated after a spinal anaesthetic showed no change The child is kept on continuous doses of Mandelamine During the year she has been under observation her general health has remained good though she does not gain weight satisfactorily There has recently been one recurrence of urinary infection of brief duration

not very dissimilar to the orifice of a diverticulum, but when the instrument is brought closer, it can be seen that the lumen of the ureter is disappearing upwards from a saccular dilatation, and the whole appearance may be likened to the old-fashioned "ear-trumpet", a convenient descriptive term differentiating it from

the "railway tunnel" orifice seen in the back pressure cases Catheterization of such an orifice is often difficult as the catheter loses its way in the saccular lower end, but in most cases a small urethroscope can be introduced directly into the orifice and its lumen followed upwards for some distance The use of this instrument makes the diagnosis from a true diverticulum a simple matter Efflux from this type of ureter is extremely sluggish, and the only movement commonly seen is a slight contraction of the ring of muscles surrounding the orifice there is a remarkable absence of the pulling upwards which often accompanies the efflux in the "railway tunnel" type While the appearance described is the most typical and the most striking, variations of it, which are probably of the same genus may be seen in older children The ureteric orifice may appear as a large and patulous looking slit, which during the efflux alters its shape to assume the "ear trumpet" appearance

(c) *Radiological* —The cystogram demonstrates a ready reflux and perfect uretero-pyelograms can be obtained in this way (Figs 75 and 76) The greater



FIG 76.—Megaureter—probably Type 5 Voiding cysto-urethrogram (lateral view) from a girl aged 7 years.

Patient had a long history of urinary infection and abdominal pains. There was no difficulty in micturition and no frequency or incontinence. The cystogram showed a large bladder which emptied without residual, and there was free reflux into very dilated ureters with comparatively normal kidneys. Dilatation was most marked in the pelvic segment of the ureters but varied considerably with the pressure in the bladder At cystoscopy the ureteric orifices were very large but retained their obliquity and both were readily dilatable. The urine was lightly infected with *B. coli* and difficult to keep sterile. There has been no deterioration in her general condition in 2 years, she is healthy growing normally and has no urinary symptoms. The appearances of the ureteric orifices in this case suggest a past infra-vesical obstruction, but there is no history to support this and she should perhaps be placed in Group 5

length of the ureter is grossly dilated and tortuous but at the lower end there is a constant relative narrowing which is best seen in the oblique views This narrow segment does not, however correspond to an apparently normal stretch of ureter as in the Type 4 megaureter but simply to the relative constriction produced by the bladder muscle and pelvic fascia. Intravenous pyelograms may show deterioration of renal function

Discussion

The explanation of these cases is conjectural There appears to be no evidence of mechanical obstruction either at the uretero-vesical level or at the bladder neck The appearances suggest a primary developmental error in the actual musculature of the whole ureter, or at least in the uretero-vesical junction, rather than any neuro-muscular imbalance The early age of the patients, the bilateral nature of the disorder and the cystoscopic appearances support this view. Our knowledge of the normal development of the musculature of the urinary tract is not adequate to allow any definite statement concerning its deficiencies It is true that while the closing membrane still occludes the uretero-vesical junction in the early embryo, a slight dilatation forms behind it, and Chwalla (1927a) has suggested that if the membrane disappeared unusually late this dilatation might result in a defective formation of muscle tissue in the mesenchyme surrounding it, thus leaving a wide open orifice and a terminal pouch to the ureter as in these Type 5 megaureters This hypothesis would indeed account for the cystoscopic appearances but must for the present remain no more than a hypothesis

Treatment and prognosis

Uncertainty as to the precise aetiology of the cases in this group makes their treatment at present symptomatic and tentative

The absence of obstruction at the uretero-vesical junction is most striking The radiography demonstrates a very free reflux on the one hand, and on the other a considerable capacity on the part of the ureter to discharge its contents on voiding There appears in fact to be a regular ebb and flow in these ureters, which means that apart from infection there is no great inherent risk to renal function We know from experience that these children can adapt to their megaureters if only they can be safe-guarded from infection Admittedly that is a big "if", so big that any reasonable operative procedure, which could be established as curative of the primary defect, would be amply justified It must be admitted that no such procedure has so far emerged. Until it does the likelihood of natural adaptation should be carefully weighed in relation to treatment and prognosis We have not found spinal anaesthesia or drugs such as mecholyl bromide of any value in these cases

The effects of infection may of course be very serious The defective ureters may be crippled by ureteritis and periureteritis, and the kidneys irreparably involved by pyelo-nephritis The onset of these complications will demand prompt specific treatment

In practice, therefore, in the early stages the first requisite is control of the infection If a sterile urine can be achieved and maintained, the child may remain in normal health, and the prospect may be regarded with some confidence. Continuous supervision is necessary so that any deterioration of renal function can be recognized at an early stage Periodic excretory pyelography is advisable so that the condition of the renal pelvis may be checked

6 **Megaureter associated with duplication**

Ureteral dilatation complicating duplication has been dealt with in the preceding chapter, which was concerned with such developmental anomalies It is well to bear in mind that a dilated ureter discovered by pyelography or cystography may

in fact be one of two and the shape of the renal pelvis should be carefully considered from this point of view In cases of unilateral megaureter the existence of a bifid ureter or pelvis on the healthy side should always suggest a similar anomaly on the other Particular attention should be paid to this point in cases of ureterocele or cystic intravesical protrusion in association with which duplications are very common. In complete pyelon duplex, where both ureters open upon the trigone, one or both may be dilated severe infection is the rule in such cases and at cystoscopy oedema frequently obscures the duplicity of the orifice

SUMMARY

When therefore, a ureteral dilatation has been recognized the first problem is one of complete diagnosis and classification. Upon this will largely depend the treatment. Some groups demand early operative relief of an obstruction at whatever level In others expectant treatment may be the wiser course provided infection can be controlled by chemotherapy and general measures

The complications of infection may render such control impossible and it is necessary to appreciate the nature and significance of such complications so that they may be dealt with as necessary

The undue length and tortuosity of the ureter may of itself determine stagnation and prevent sterilization of the urine. Fixation of the loops by periureteral adhesions intensifies the condition The indication is to expose the ureter free it and excise any redundant length So long as drainage to the bladder is satisfactory this may be a valuable procedure.

The pelvi ureteric junction may be caught up in inflammatory adhesions causing a hydro-nephrosis (*see* page 137) The renal pelvis becomes a source of persisting infection and grave pyelo-nephritis soon follows An early exposure of the kidney adhesiolysis and if necessary drainage of the pelvis is urgently called for Stone is a serious complication in a megaureter particularly in Type 4 of our classification Removal of the stone even with the fullest possible dilatation of the uretero-vesical opening, is very likely to be followed by recurrence Excision of the uretero-vesical orifice and re implantation of the ureter so as to ensure a free reflux offers the best hope of any further crystals being flushed clear but in unilateral cases nephro-ureterectomy may be necessary

Contra-lateral infection

In a unilateral dilatation there is always a risk of infection spreading to the opposite ureter The possibility should deter us from persisting over long with conservative measures. In severe cases, especially with a kidney known to be damaged, total extirpation should be carried out without hesitation.

CHAPTER 10

URINARY OBSTRUCTION—HYDRO-NEPHROSIS

DILATATION of the renal pelvis (hydro-nephrosis) is met with in many circumstances and in varying degrees

Distensibility of the pelvis

The normal renal pelvis appears to be less distensile than the ureter We should expect this to be so since any undue increase in intrapelvic pressure must exert a prompt retarding effect upon the renal circulation to compensate Relief of hydro-nephrosis is by so much the more urgent if renal damage is to be prevented

It is rare to see any bulging of the pelvis in normal pyelograms· when persistently present, even in slight degree, it must be considered abnormal, possibly due to atony of the wall or dysfunction of the pelvi-ureteric junction.

BACK-PRESSURE HYDRO-NEPHROSIS

Hydro-nephrosis sooner or later ensues upon dilatation of the lower pyelon segment (megaureter) as a back-pressure effect, when it is often bilateral In such cases pyelographic evidence of pelvic distension is clearly an ominous sign, especially if it seems to be increasing, relief of the causal obstruction becomes the more urgent

A specific lesion such as a calculus or neoplasm may partially fill the pelvis, blocking its outlet In such cases calyceal dilatation is likely to occur early and the pyelogram may show the pelvis itself relatively less dilated In the idiopathic form of hydro-nephrosis the reverse is usually true, and calyceal dilatation indicates a block of long standing.

Clubbing of the calyces alone may occasionally be seen, possibly due to dysfunction of the muscle of Muschat

Inflammatory adhesions

In infected megaureters (and possibly also in some cases of pyelitis), as stated in Chapter 9, the upper ureter may become kinked and the pelvi-ureteric junction fixed and occluded by inflammatory adhesions This results in obstructive hydro-nephrosis and persisting infection until relieved by adhesiolysis In such cases the adhesions are typically inflammatory and recent in type and, when freed, the pelvi-ureteric junction appears perfectly normal

HYDRO-CALYCOSIS

Localized dilatation of a single calyx (hydro-calycosis) may result from blockage following infection or, for example, by a stone

ACUTE INFECTIVE HYDRO-NEPHROSIS

Acute distension of the pelvis may occur in pyelo-nephritis and is recognized by unduly severe local pain, rigidity and tenderness The tense kidney is usually palpable. In such cases the bladder urine may be clear The kidney as a rule drains naturally within 24 hours or so the emergency passing and the bladder urine becoming purulent. but in some cases it may become advisable to pass a ureteric catheter to drain the pelvis. On rare occasions a pyonephrosis may ensue (*see* Chapter 5) Generally speaking, however the effects of pyelo-nephritis upon the pelvis as seen in pyelograms, are transient fuzziness or later contraction rather than distension.

CONGENITAL OR IDIOPATHIC HYDRO-NEPHROSIS

There remains the large group of hydro-nephroses with which we are now concerned in which the cause is less easily identifiable In this type sometimes termed congenital or idiopathic the obstruction at the pelvi ureteric junction is variously ascribed to kinks and adhesions to stricture, to aberrant vessels or occasionally to achalasia The lesion is most often unilateral but bilateral instances are seen from time to time

Pathology

The dilatation affects chiefly the pelvis and as has been stated, dilatation of the calyces at the expense of the kidney substance occurs later in the course of the disease The effect of the obstruction upon the musculature of the pelvis is to cause hypertrophy with an increase of the elastic tissue and later on a thinning and weakening of the pelvic wall. The nephrons are damaged by the back pressure both directly and indirectly by interference with their blood supply At first it seems that re-absorption from the pelvis helps to maintain renal function but there must be an early limit to this possibility The subsequent fate of the kidney substance depends upon the degree and constancy of the obstruction and upon the presence or absence of infection Where more complete obstruction has occurred early in life, the kidney may be reduced to a thin shell of tissue upon a huge hydro-nephrotic sac, symptomless except for its mass. In less complete obstructions a considerable degree of renal function may be maintained for many years but deterioration rapidly ensues if the hydro-nephrosis becomes infected As with stasis elsewhere in the urinary tract, stone formation is apt to complicate the condition and a group of small sterile stones may be found in a dilated lower calyx, though this is rare

The pelvi ureteric junction

A more detailed examination of the pathology at the pelvi ureteric junction is an essential preliminary to a discussion of conservative surgery The most common finding in our series has been a kinking of the upper ureter which is bound down to a distended pelvis in the manner illustrated in Fig. 77 The adhesion which binds it is sometimes a loose sail shaped fold sometimes much tighter and with it is often associated a small vessel in an aberrant position (as described by Ostling, 1942) Occasionally the ureter is actually running intramurally within the muscular coat of the pelvis and the kink can only be straightened by stripping the ureter

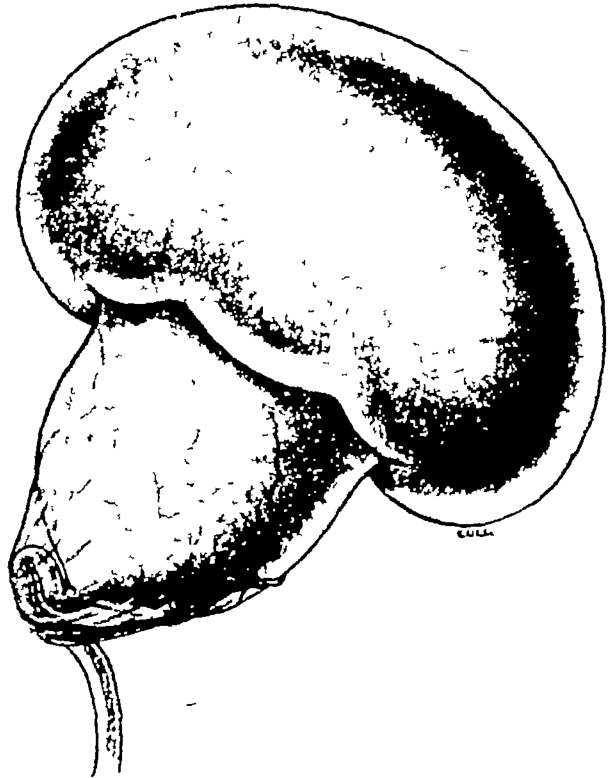


FIG 77 —Hydro-nephrosis Diagrammatic drawing of the "sail" adhesion As described in the text, the kinked ureter in many cases is more intimately fused with the wall of the pelvis than is here indicated



FIG 78 —Diagrammatic drawing to illustrate gap in the muscular wall of the ureter

down to its mucosa In the latter instance there can be very little doubt that the kinking is congenital in origin in other cases it is not so certain but these adhesions can hardly be the result of inflammation as the great majority of those showing them have sterile urine and no history of an infection It has been suggested that the kinking is simply the effect of the distension of the renal pelvis which has occurred because of some other lesion and there can be no doubt that the kink is occasionally associated with a definite narrowing immediately below the junction of pelvis and ureter If there should be a temporary distension of the pelvis, due for instance to a diuresis, there is no doubt that the formation of a kink would accentuate and perhaps perpetuate the obstruction at the pelvi ureteric junction which was previously no more than a relative narrowing. The stricture found at this point alone or in association with a kink is seldom a fibrous stricture but merely a narrow muscular segment (*see* Fig 78) This again may be of congenital origin since in this area tight folds are found in the wall of the ureter during late foetal life (*see* Chapter 6) accompanied by some abnormality in the muscular layer The blood vessels which play a part in the pelvi ureteric obstruction are most often arteries running to the lower pole of the kidney either from the renal artery or directly from the aorta they are usually placed behind the ureter though they may be in front and on occasion the vessel is a vein Not infrequently the aberrant vessel is associated with the typical kink and adhesion already described and perhaps on these occasions the pelvis as it distends has pushed itself forward between the vessels of the pedicle so that the vessel itself is an accessory factor only There are a few cases (rare in our series) in which the pelvi ureteric junction seems entirely normal and the cause of the distension is obscure It may be that the upper segment of the ureter fails to relax in co-ordination with the contractions of the pelvis (achalasia) but there is very little convincing evidence for this.

Associated changes in the ureter

It is common in children to see the ureter below a hydro-nephrosis dilated The dilatation affects chiefly the lumbar segment and is presumably a disuse effect (slacking ureter) in the sterile cases When infection is present this may also be a factor inducing atony

Infection

Many hydro-nephroses remain sterile infection when it occurs, is always a disquieting complication If transient the effects may not be unduly serious but if persistent, deterioration of function is inevitably accelerated. In severe suppura tive form pyonephrosis spells total loss (*see* Chapter 5)

Clinical features

Hydro-nephrosis of the extreme type commonly seen in infancy presents as a symptomless tumour filling the loin and possibly the whole abdomen When very large it may be impossible by clinical examination alone to determine the side of origin indeed an ovarian or mesenteric cyst may be simulated One can usually note that the outline of the tumour is smooth and rounded If very tense a solid tumour may be simulated, in particular a renal blastoma, but an even contour is very unusual in solid growths, and a hydro-nephrotic sac usually trans-illuminates well a simple observation which may well settle the diagnosis forthwith. The excretion pyelogram if obtainable will show total absence of secretion on the

affected side, which is rare in renal embryoma (*see* Chapter 13) If the other kidney is shown by pyelography to be normal, cystoscopy is unnecessary, but if it is carried out no efflux is seen on the affected side The ureteric catheter is usually checked at the pelvi-ureteric level, but in some cases it enters and drains the contents of the hydro-nephrosis In such cases the sac may be decompressed as desired

Treatment of this type of hydro-nephrosis is inevitably nephrectomy. The loin incision is always preferable Even the largest of sacs are best dealt with by this route, any mechanical difficulty from size being got over by aspiration

Most cases of hydro-nephrosis as seen in childhood are, however, fortunately not of this extreme degree, and the majority are by no means symptomless

A sterile hydro-nephrosis in the evolving stage gives rise to periodic attacks of pain and vomiting of striking character, but in the child they are often misinterpreted The pain and vomiting are severe, lasting for some hours, and the child is restless, miserable, somewhat shocked and compelled to lie down Some of these children have learnt the trick of relieving their discomfort by posture, a fact which may be of assistance in diagnosis During the attack the kidney may be palpably enlarged and tender, going down rapidly again with subsidence of the pain Older children will usually indicate the loin as the site of their pain, but the younger ones are apt to be vague the attack is easily mistaken for an abdominal crisis and a misguided appendicectomy performed. Numbers of our children have a history of attacks of this kind extending over several years before their true nature has been suspected This is tragic, for conservative surgery can save many of these kidneys if they are recognized in time The attacks are often followed by the passage of surprising amounts of urine, which may be out of all proportion to the apparent size of the dilatation as shown pyelographically, for example, a boy of 10 years passed 45 ounces in the few hours following an attack of pain

The infected hydro-nephrosis tends to give a different clinical picture. The presenting feature is usually one of persisting infection Pain is less prominent and the brisk attacks described above are not seen as a rule In these instances the hydro-nephrosis is discovered by routine pyelography and its size may be surprising in the absence of pain or palpable enlargement of the kidney It seems probable that the dilatation of the pelvis in this form of hydro-nephrosis is largely due to atony of the wall, the result of infection of the same nature as the ureteral "ileus" (*see* Chapter 9)

Clinical signs

On physical examination, the hydro-nephrotic kidney is palpable in a proportion of cases In the acute phase it may present as a considerable tense tender loin swelling The majority of cases, however, are not seen under such favourable circumstances In some, one can detect the enlarged kidney simply as such Now and then a soft cystic swelling can be recognized, especially by comparison with the opposite loin In many no palpable swelling can be recognized and the diagnosis depends entirely upon pyelography

Urine

Complete urinalysis, chemical and bacteriological, is made If infected, the sensitivity of the organisms must guide essential chemotherapy



FIG 79.—Hydro-nephrosis (left) Retrograde pyelogram from an infant girl aged 1 year an eczematous baby who suffered from abdominal pain and persisting urinary infection.

At operation on the left kidney an aberrant vessel was found constricting the pelvi-ureteric junction, this was ligatured, the pelvis opened and the ureter dilated. Two years later there had been no further symptoms, the urine was clear and an intravenous pyelogram showed the kidney secreting well, though some dilatation persisted. Incidentally the child eczema cleared very rapidly after the operation on the kidney



FIG 80.—Hydro-nephrosis. Retrograde pyelogram (note kinking of the proximal ureter) from a boy aged 8½ years.

At operation division of the ureter and re-implantation was necessary. One month later excretion pyelogram showed the left kidney secreting well and the ureter filling satisfactorily. The boy has remained symptom-free (3 years).

The shadow shown on the right side is a calcified tuberculous gland.

(*a*)



FIG 81 —Hydro-nephrosis (left) Excretion pyelograms from a girl aged 12 years (*a*) Before operation, (*b*) two years later, showing pelvis comparatively normal and ureter filling well

(*b*)



Further investigations

Pyelography

This affords evidence which is essential not only to diagnosis but to prognosis and treatment. In unilateral cases there is the useful contrast of the sound kidney The characteristic finding in an early case is a bulging convex pelvis, tending to be spherical in outline and sharply cut off at or a little below the pelvi ureteric junction (Fig. 79) Where there is a kink or a vessel a short segment of the upper ureter may be filled and is distorted, often directed laterally (*see* Fig 80) The calyces show a little blunting at first, but with the progress of the disease become grossly dilated, and where the function of the kidney is poor the pyelogram may show no more than a series of blobs in the periphery of an enlarged kidney Later still there may be no function detectable by radiology though this does not necessarily imply that function is irrecoverable.

Cystoscopy

This is usually essential to determine the condition of the ureter to verify if need be the extent of the hydro-nephrosis and if possible, to obtain a specimen of its contents. In the case of a pyelographically silent hydro-nephrosis it may be essential to determine that a kidney is in fact present.

Treatment

The ideal treatment of this type of hydro-nephrosis is to relieve the obstruction and preserve the kidney In childhood this is possible in a high proportion of cases seen (approximately 60 per cent of our last 70 cases) In the most favourable restoration of function as shown by post-operative pyelography is virtually complete (*see* Fig. 81 (*a*) and (*b*)) In the less fortunate the relief of symptoms, the clearing of infection if present, and the considerable degree of renal function regained, justifies conservative surgery There is no evidence in our cases, some of which have been followed into adult life that the subnormal kidney tends to a hyperpiesia. All our cases have kept a normal blood pressure following operation In one a pre-operative hypertension persisted and was only relieved by a secondary nephrectomy (*see* Chapter 4 Renal Hypertension, case C H)

The pre-operative investigations in particular pyelography give a clue to the feasibility of a conservative procedure

Evidence of low-grade function is naturally a deterrent to conservatism, especially in the presence of infection

The final decision however not infrequently depends upon the actual condition of the kidney as found at operation Chronic pyelo-nephritis, shown by scarring and hardness of texture is an unfavourable sign If it is marked, nephrectomy is probably the wiser course. When the kidney is natural in colour without much evidence of sclerosis, even though it be soft and flabby from calyceal dilatation it is worth preserving There are many times when the decision is difficult, especially in bilateral cases If in serious doubt, nephrectomy should be done Increasing experience, however has taught us that more kidneys appear conservable than we used to think.

The conservative operation

Preliminaries

In infected cases an attempt should be made to sterilize the urine. The blood should be examined, the grouping should be known and, if it seems necessary, a drip transfusion is set up some hours before the operation and may be continued into the immediate post-operative period. In practice, however, transfusions are not often needed The operation, though it may be tedious, is seldom attended by much shock and most of our cases show surprisingly little reaction

The incision should be ample enough to facilitate easy delivery of the kidney and adequate exposure of the upper ureter If need be the last rib may be divided and removed, though this is rarely necessary The kidney is gently cleared of its perirenal fat and brought well out into the wound It is held forwards by an assistant, the renal bed being loosely filled by a narrow gauze pack Haemostasis throughout must be meticulous The kidney is now inspected, its condition estimated and the decision made as to whether or not it is worth conserving If the kidney does appear reasonably good, the hydro-nephrotic sac and pelvi-ureteric junction are carefully cleared by gentle dissection. It is helpful to isolate the ureter below the junction and pass a length of tape beneath it to act as a retractor The condition at the pelvi-ureteric junction is thus exposed to view and the requisite further steps determined In the simplest cases separation of adhesions alone may suffice This applies mainly to the secondary inflammatory adhesions already referred to (page 154), but in some of those which are congenital (the "sail" adhesion), the kinked adherent ureter proves easily separable from the pelvis and, once freed, the pelvis is seen to empty rapidly. The obstruction is clearly rectified and nothing further is necessary In other cases, the ureter is so embedded that, when finally freed, the wall is left seriously denuded and re-implantation is necessary. On full exposure the pelvi-ureteric junction may be found unduly narrow. It is not always easy to be sure about this, the dilatation of the pelvis above and of the ureter below tends to make the appearance deceptive. If in doubt, the pelvis should be opened and bougies passed from above downwards When there is genuine narrowing it seldom appears to be a fibrous stricture One may feel satisfied that simple dilatation is enough If, however, enlargement of the outlet is clearly required, some form of plastic procedure must be adopted We have employed one or other of the following methods

(1) The simpler, by incising the whole thickness of pelvis and ureter through the junction in a vertical direction for about $\frac{1}{4}$ inch and suturing the wound transversely with a few interrupted sutures of fine unhardened cat-gut passed atraumatically on the smallest possible needle This manoeuvre has proved very satisfactory in many of our cases On occasion a "Y"–"V" plastic procedure as suggested by Foley (1937) has been found useful

(2) The ureter is divided and re-implanted Care is necessary in the child's ureter to ensure an adequate lumen. The anastomosis should be made very obliquely, or the cut upper end of the ureter may be nicked on either side, and the tiny flaps so attached to the pelvis that they are held widely open

In all cases a ureteric catheter, brought out through the wound, is left in

position for 48 hours as a splint to the anastomosis, and the pelvis is also by a small soft rubber catheter *

In a large hydro-nephrosis partial pelvectomy is usually carried out a closure by interrupted infolding sutures of unhardened cat gut

The high pelvi-ureteric junction

In rare cases the ureter is found to join the pelvis much above its normal p thus contributing to hydro-nephrosis In such cases re implantation is ol necessary We have had one such case which has been quite successful.

Vascular obstruction

Whatever may be the exact evolution of this variety of obstruction it enough at operation that the aberrant vessel and its associated ba maintaining the obstruction and the pelvis and ureter must be freed if th is to be relieved

In a number of cases the sail adhesion is seen with only a tiny vessel ated, and in these it is clearly the adhesion which is responsible for the obst Even when a large vessel is found to be tightly compressing the ureter it worthy that there is, as a rule a length of ureter above the vessel whic dilated but is flattened and fixed by the adhesions The tethering bands important factor in vascular obstruction and to divide and free them tho is essential

In our earlier cases the procedure adopted was to divide the vessel an artery after having dissected away all tethering adhesions The im collapse of the pelvis and filling of the ureter is most striking and the afte have been invariably excellent. Immediately following division of the ve associated segment of the kidney darkens in colour No post-operative hae has been seen in our cases, but no doubt some degree of renal atrophy mus

In recent cases we have carried out the manoeuvre which we owe to H Stewart (1947) of folding up the kidney so that the vessel rides upwards pelvis thus releasing the obstruction. We have been most impressed efficacy

*The Genito-Urinary Manufacturing Co Ltd. now makes for us a modified Malecot A fine soft-rubber extension of suitable size is fused to the tip of the Malecot the exten is passed down the ureter through the junction the head of the Malecot resting w pelvis. The one tube then serves both for splintage and drainage.

CHAPTER 11

UROLITHIASIS

LITHIASIS is no longer a common disease of childhood in Great Britain, and in the majority of cases seen today the stone itself is secondary to infection or stasis in the urinary tract The therapeutic problem is correspondingly a complex one, simple removal of the stone is not enough and if recurrence is to be avoided careful attention must be paid to the underlying pathology and biochemistry

Composition and structure

The classical descriptions of the disease have familiarized us with certain characteristic stones the features of which must be briefly recalled

Calcium oxalate stones are formed in a sterile acid urine, they rarely attain any great dimensions, in the kidney and ureter they are found as small smooth reddish-brown stones, or covered with sharp glistening crystals and darkened by the blood pigment In the bladder they appear as mulberry calculi or "jack stones"

Calcium phosphate may occur in a crystalline form (apatite), often in conjunction with some calcium carbonate These crystalline phosphate stones can be formed in sterile urine, but much more commonly amorphous calcium phosphate is laid down in infected strongly alkaline urine. Such stones are often large, they are laminated, with a friable surface and a muddy yellow colour In the renal pelvis they frequently assume a staghorn appearance Ammonium magnesium phosphate occasionally occurs alone but more often in conjunction with amorphous calcium phosphate in urine infected with urea-splitting organisms

Uric acid and ammonium urate form rather smooth red-brown stones in sterile acid urine, they are much more commonly found in the bladder than in the upper urinary tract

Cystine stones have a greenish-yellow, smooth, almost waxy appearance and are formed in acid urines In the bladder they are usually spherical or ellipsoid, in the kidney frequently dendritic Conglomerate stones are sometimes seen in which a number of small elements have fused together to form a large stone

It must be emphasized, however, that the majority of stones are of mixed composition and that oxalates and urates are frequently found in typical phosphate stones, moreover, since the majority of cases in children are infected, phosphates predominate

A calculus is not simply a crystalline structure but contains a variable amount of colloidal organic matter In most cases a definite nucleus can be demonstrated around which a series of laminae have been deposited The lamination is due to alternate layers of closely packed crystals with little supporting colloid, and spongy layers containing much albumin In the infected cases the nucleus may consist of a lump of pus, a blood clot, slough or foreign body Hellstrom (1936) has

particularly emphasized the frequency with which masses of staphylococci occur in the nucleus of infected stones

The nucleus of the sterile primary stones has been studied in some detail in the hope that it would throw some light upon the pathogenesis Randall (1940) has described calcareous plaques which can be found beneath the mucosa of the renal papillae and calcareous deposits in the collecting tubules in his opinion primary stones are formed upon a nucleus of one of these plaques which has sloughed through the over lying mucosa, so that the stones are at first adherent to the papillae However plaques were found in 19 6 per cent of all *post mortems* (at all ages but most frequently in middle life) and it does appear that some other factor must be required in order to form a stone or the disease would be a great deal more common than it is Hammarsten (1945) has studied the formation of nuclei in experimental oxalate stones and in sulphonamide concretions it appears that these substances, when they reach a certain concentration separate out as tiny globules (micelles) resembling drops of oil These probably consist of very concentrated colloidal solutions but in time crystallization takes place within them Prien (1949) in a careful crystallographic study of the nuclei finds evidence that stones may be formed in either of these ways

Aetiology

There can be no doubt that calculous disease has changed its character since it has become less frequent. in the stone areas, such as Norfolk and Derbyshire during the last century as in India and China today stones were common in children usually vesical, and uric acid and oxalates predominated being formed in a sterile urine It is almost certain that a dietary factor was responsible for the endemic type of calculus, not of course a simple matter of excessive intake of stone forming substances (indeed urate and oxalate compounds are almost absent from the diet of a Chinese coolie (Jolv 1929)) but some more complex defect possibly a vitamin A deficiency (McCarrison 1927) An almost exclusively cereal diet has been a common finding in the stone areas Lithiasis is not often seen where milk and butter are plentiful

Stone forming substances

No simple dietary explanation can be given for the cases of calculous disease now to be observed in European children which are usually septic and found in the upper urinary tract Naturally considerable attention has been paid to the meta bolism of the stone forming substances Uric acid is derived from the breakdown of nucleo-protein and may be endogenous or exogenous Uric acid is found in somewhat greater concentrations in the urine of young infants and may form infarcts in the kidney of the new born it is difficult to link either of these facts to the formation of calculi since stones are seldom observed within the first 6 months and uric acid stones are now uncommon in children of any age In later life attempts have been made to correlate uric acid calculi with an excessive indulgence in meat and with the renal uric acid deposits in gout, but again the link has not been established Oxalate may also be derived from endogenous or exogenous sources rhubarb spinach, asparagus and other fruits being notoriously rich in the substance. Oxalate excretion does not cease on an oxalate-free diet, and Hammarsten

1937) has even been able to produce oxalate calculi experimentally under these circumstances The passage of oxalate crystals is not uncommon, but does not necessarily indicate an excessive excretion and has little relation to the formation of stones The crystals passed in oxaluria are the dihydrate, whereas stones are almost always composed of the monohydrate crystal Both oxalates and urates are held in urine in super-saturated solution, that is to say, urine contains about 10 times as much uric acid as could be contained in a similar quantity of water. This super-saturation is probably accounted for by the adsorption of the inorganic material upon the surface of colloidal particles Schade (quoted by Joly, 1929) suggested that some breakdown of the protective action of the colloids might be responsible for the formation of calculi unfortunately this hypothesis has not proved very helpful, the fact that comparatively pure stones occur argues against such a non-specific process, and in conditions of stasis and sepsis, in which stones are prone to form, the colloids are considerably increased

Phosphates occur in food either as inorganic salts, the absorption of which is largely influenced by the *p*H of the intestinal contents, or in complex organic substances Urinary phosphates are increased on a high protein diet, but the element cannot of course be eliminated from the urine Phosphaturia is evidence of alkalinity rather than increased excretion Ammonium magnesium phosphate is not normally found in the urine in any significant quantity but is produced by the action of urea-splitting organisms

Calcium excretion

In recent years investigators have paid less attention to the acid radicals and more to the calcium element It is now clear that where calcium excretion is excessive, as for instance in hyperparathyroidism, calcium stones are formed but they may be oxalate or phosphate or a mixture Calcium is normally excreted very largely by the bowel and urinary values are low Hammarsten (1937) has shown that when magnesium is given in adequate quantities very little calcium is found in the urine, on a diet poor in magnesium, on the other hand, urinary calcium rises and stones may be formed, particularly as the magnesium has also some action in maintaining the oxalates in solution Curiously enough this worker also found that decreasing the calcium in the diet led to a mobilization of that element from the bones and to increased urinary calcium Citric acid also has some influence in alkaline urine soluble calcium citrate tends to replace the insoluble calcium phosphate and in certain cases of recurrent stone formation without infection the urinary citrates have been shown to be abnormally low

Increased calcium excretion is of the greatest importance in the aetiology of calculi and the conditions in which it occurs must be discussed in some detail Flocks (1939) found that over half his cases of calculi had a high urinary calcium, although no abnormalities could be found in the blood Albright and Reifenstein (1948) have called this condition, described by Flocks, idiopathic hypercalcuria, but there is very little confirmatory evidence and it is hard to assess the significance of Flock's observation It is interesting to notice that the urinary excretion of calcium was raised even further when these patients were put upon an acid ash diet, whereas in the cases with normal calcium excretion this diet had the reverse effect

Hyperparathyroidism

Hyperparathyroidism is uncommon in children but deserves mention because it has formed the starting point for so much research on calcium metabolism The first effect of parathormone excess is apparently to increase the renal excretion of phosphates and thus depress the serum phosphorus values by some unknown mechanism this causes the mobilization of calcium from the skeleton and an increase in the serum calcium Finally the serum increase produces a urinary calcium increase The effects upon the kidney are threefold (*i*) polyuria necessary to ensure excretion of large quantities of the salts, or due to renal damage (*ii*) nephrocalcinosis—deposition of calcium within the substance of the kidney chiefly in the region of the distal tubes and (*iii*) nephrolithiasis the stones are always calcium ones in the first place though they may be oxalate or phosphate (Keating and Cook, 1945 found 6 out of 9 to be oxalate) but when infection has supervened ammonium magnesium phosphate is also formed

There has been considerable dispute as to the frequency with which hyper parathyroidism presents with urinary calculi as its only sign Keating and Cook estimate that 2 per cent of stone cases are due to this disease The diagnosis is not always easy to establish and a full investigation demands calcium balance estima tions which cannot be made in many hospitals probably the safest rule is that all stone cases should have the Sulkowitch test performed (Barney and Sulkowitch 1937) and if this reveals abnormally high urinary calcium the serum values should be estimated Raising of the serum calcium (normal 10 mg per 100 ml $\pm$ 0 5) is a more constant finding than lowering of the serum phosphorus which may be normal or even raised if the renal damage is severe The alkaline phosphatase is not raised in cases with renal disease but without skeletal involvement

Immobilization

Urinary calculi are common in patients with extensive injury or disease of bone particularly when they are immobilized in the recumbent position The bone disease itself or merely the disuse causes a decalcification of the skeleton and calcium is excreted in the urine in large amounts In adults the serum calcium is scarcely ever raised under these circumstances but in children it may be Albright and Reifenstein have reported such a case a child with osteomyelitis and urinary calculi Recumbency itself adds to the likelihood of stone formation by causing stagnation in all the calyces so that any crystals which appear tend to remain in the kidney instead of passing down the urinary tract. Calculus has been particu larly common in children who have been exposed to long periods of direct sun light while being immobilized for the treatment of bone and joint tuberculosis (Mawson 1932) and it is possible that oliguria due to slight dehydration may have added to the effects of recumbency and decalcification in these cases The typical recumbency calculus is a soft rapidly formed calcium phosphate one often symptomless until the patient gets out of bed or showing itself only by urinary infection or haematuria A few cases of spontaneous resolution of calcium shadows within the urinary tract have been described but more often dendritic calculi are formed which defy conservative treatment.

Osteoporosis with excessive calcium excretion and consequent urinary calculi have been reported in Cushing s syndrome and hyperthyroidism Stones are also described in patients who have been given excessive doses of vitamin D

Nephrocalcinosis

Biochemical disorders involving calcium metabolism may be due to primary renal pathology. This appears to be the case in the nephrocalcinosis associated with hyperchloraemic acidosis, a disease occurring both in infants and in adult life, in which radiological examinations show a stippled calcification within the kidney, chiefly in the area of the renal pyramids. There is a severe and persistent acidosis despite a fixed urinary alkalinity and polyuria The disorder has been regarded as the result of a failure on the part of the distal convoluted tubules to manufacture ammonia, with consequent loss of fixed base in the urine, but recent evidence (Latner and Burnard, 1950) suggests that the defect is actually situated in the proximal tubules, which fail to re-absorb bicarbonate, thus overtaxing the capacity of the distal tubules to adjust the *p*H of the urine As Albright and Reifenstein have shown, the acidosis may cause mobilization of the calcium reserve in the bones, and a type of osteomalacia Such patients are greatly improved by the ingestion of a citric acid–sodium citrate mixture which, since the organic acid radical is metabolized, leaves an excess of sodium to neutralize the acidosis It is not clear whether the calcification within the kidney is primary or merely the result of the biochemical disorder. probably the latter, since Albright and Reifenstein found no further calcification occurring while patients were on citrate treatment. The association of calculi with nephrocalcinosis has been recorded in several cases, but not in the infantile group the urine is in most cases sterile, but severe infections may occur Occasional cases are encountered in which the radiological examination shows the typical nephrocalcinosis with stones, but in which no biochemical disturbance can be discovered Davis, Klingberg and Stowell (1950) have reported a case of nephrocalcinosis in a boy of 12 years who passed oxalate crystals and was found *post mortem* to have similar crystals within the bone marrow

Calcification within the renal parenchyma may occur in a wide variety of other conditions but seldom results in the formation of urinary calculi Thus in hypervitaminosis D, calcification and renal failure are not infrequently encountered, similarly the aftermath of mercurial or sulphonamide renal damage may involve the deposition of calcium Calcification is well recognized in tuberculosis and neoplasm, but the differentiation of these diseases from calculus formation rarely presents any difficulty

Cystine

Cystine is not normally found in the urine and cystine stones are only found in patients with a hereditary disorder known as cystinuria These patients excrete a fairly constant amount of cystine throughout life, either in solution or in the form of crystals Cystinuria has been regarded as an inborn error of metabolism, but the work of Dent (1950) suggests that the error is in the renal tubules, blood values for cystine appear to be normal but the renal threshold is low and lysine and arginine, which have a similar molecular structure are constantly present in the urine as well as cystine The condition may well be comparable to renal glycosuria Only a small percentage of cystinurics develop stones, Joly (1929) estimates only 2·7 per cent, and the precipitating factor in stone formation is not clear. Calculi are laid down in sterile acid urine and there appears to be a slight tendency

to spontaneous solution in strongly alkaline urine The passage of crystals is not related to calculous disease. Cystine cannot be eliminated from the urine of cystinurics as it is partly endogenous in origin but excretion is increased by a heavy protein diet especially by the ingestion of cysteine and methionine and other sulphur-containing amino acids Cystine itself does not appear to be absorbed from the intestine in significant quantities

Cystinuria must be distinguished from cystinosis in which cystine crystals are deposited in many parts of the body though stone formation is uncommon Cystinosis occurs in children with Fanconi s syndrome (a vitamin D resistant type of rickets) in which all the amino acids as well as sugar are present in the urine (McCune Mason and Clarke, 1943) Although a disorder of the renal tubules may again be the fundamental pathology the metabolic disturbance is the most important feature in these cases and the urologist is unlikely to be concerned in the treatment

Age and sex incidence

Winkel-Smith (1944) notes that two-thirds of his patients developed their calculi before they were 5 years old in the Great Ormond Street series symptoms appear to have begun most often between 6 months and 3 years of age, but as in some cases it is certain that the infection was present before the stone it is hard to give an accurate estimate of the age at which the stone formed We have noted that the formation of stones demonstrable radiographically may occur with surprising rapidity (2–3 months)

In the vesical calculus of endemic lithiasis males were far more often affected in the infected upper tract type of stone now seen, this sex difference is not so pronounced, but about 60 per cent of our cases have been boys

Clinical and pathological types

It is not proposed to mention here all the possible variations of calculous disease but simply to draw attention to the characteristic syndromes found in children Approximately 90 per cent of cases attending Great Ormond Street with stone have had infected urine and in the majority of cases pyuria has been evident to the naked eye Pain was the presenting symptom in approximately one third of the cases From these two facts alone it will be clear that the small stones confined to the calyces, and the small stones which pass down the ureter during attacks of renal colic, are very much less frequent in the child than in the adult. It is perhaps in the behaviour of the ureter that the child differs most from the adult the child s ureter is readily dilated and allows stones to pass on to the bladder unless it is itself already abnormal

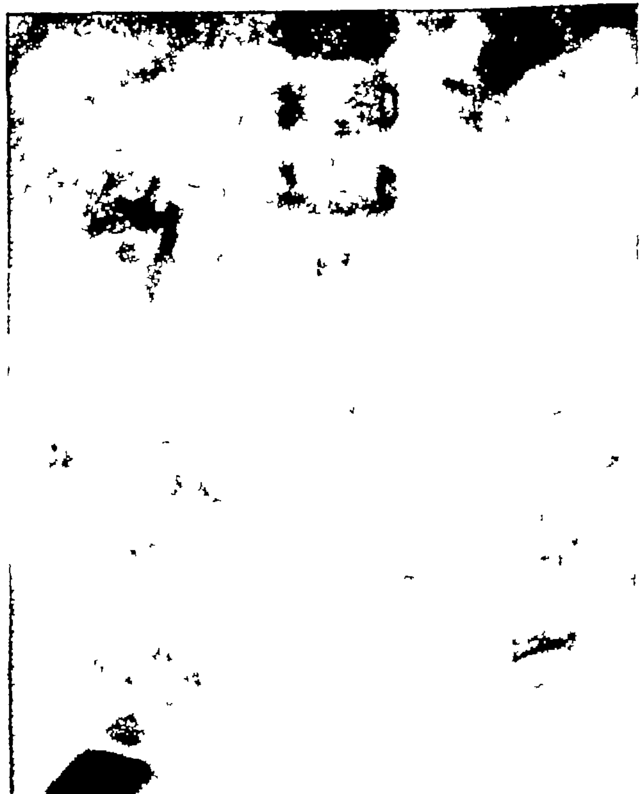
Stone in megaureter

Renal and ureteric calculi are often found in association with the type of megaureter unilateral or bilateral, which has an apparently normal or a slightly stenosed lower end (*see* Chapter 9) and which in these cases is commonly infected with *B proteus* The stones are large oval laminated phosphate stones Often 2–3 may be seen one on top of the other in the distended pelvic segment of the ureter and there may in addition be dendritic calculi in the kidney (*see* Fig. 82 (*a*) and (*b*))



(*a*)

FIG 82 —Ureteric calculi with megaureter (bilateral) Boy aged 1 year 7 months (*a*) Skiagram showing calculi lower ends of both ureters (one in the urethra) and a further collection in the right kidney, (*b*) intravenous pyelogram after removal of ureteric stones, showing bilateral megaureter and remaining calculi in lower pole of right kidney



This little boy had suffered from persisting nary infection and had passed a stone per thram Right and left uretero-lithotomy l right nephro-lithotomy were performed stages and the urine, heavily infected h *B proteus* was ultimately cleared with eptomycin Six months later an intravenous elogram showed good renal function on h sides The lower end of both ureters nained markedly dilated Unfortunately infection returned and further calculi med in the lower pole of the right kidney se increased in size rapidly (6 months) uming a dendritic form and filling the vis In view of the persisting infection the ht kidney was removed and the urine then came sterile At a later date further stones formed in the left ureter and these were noved since when progress has been isfactory (Calculi phosphatic)

The type of megaureter which has a wide gaping ureteric orifice does not predispose to stone formation probably the flushing action of the reflux at each act of micturition is sufficient to remove any debris likely to form the nucleus of a stone.

The symptoms of this type of stone are little more than the symptoms of the infected megaureter—pyuria recurrent pyrexia haematuria and dull pain in the loin Stones may also occur in a ureterocele and at times the dilatation of the ureter is secondary to the impaction of a stone at the lower end though we believe this to be less common than primary dilatation

Staghorn calculi

This type of stone is common in children and in some of our cases a severe pyelitis was known to have preceded stone formation Again *B proteus* is the common infecting organism and again it is the pyuria and recurrent fever rather than any specific symptoms which draw attention to the stone. The renal function in such cases is often well preserved and the stone may cause surprisingly little dilatation of the calyces a feature which does not assist the surgeon in removing it. These calculi must always be associated with inflammation and ulceration of the pelvic mucosa which together with the resistant infection are responsible for the high rate of recurrence after lithotomy

Impacted pelvic stones

Pain is a more constant feature of the calculous hydro-nephrosis due to impaction of a stone at the pelvi ureteric outlet. There is comparatively rapid deterioration of renal function and stagnation in the distended calyces is often responsible for small secondary stones formed either before or after removal of the pelvic stone Dilatation of the lumbar spindle of the ureter without obstruction (inflammatory ureterectasis *see* Chapter 9) is seen in association with either type of stone in the renal pelvis and usually subsides after lithotomy (*see* Fig. 83 (*a*) and (*b*))

Stone in congenital pelvic hydro-nephrosis

Calculi occasionally complicate hydro-nephrosis due to bands aberrant vessels and stricture at the pelvi ureteric junction They are presumably the result of stasis and may occur in sterile urine The symptoms are those of hydro-nephrosis (that is severe attacks of pain in the loin with vomiting) but the stones themselves may be responsible for attacks of haematuria (Fig 84 (*a*) and (*b*)) Where infection has supervened, the symptoms are those of a pyonephrosis

Stones in anomalous kidneys

Stones are not infrequently found in kidneys with a double pelvis and ureter the lower and larger pelvis being more often affected, but these stones do not differ in type from those found in normal kidneys Stones may also occur in fused kidneys, presumably the result of stasis in the abnormally formed pelves (*See* Appendix III case 4)

Vesical calculi

All cases of vesical calculi treated in the last 12 years at Great Ormond Street have been associated with upper tract calculi or with foreign bodies in the bladder All have been infected and phosphates have formed the greater part of their structure. The symptoms have been either those of pyuria and recurrent infection



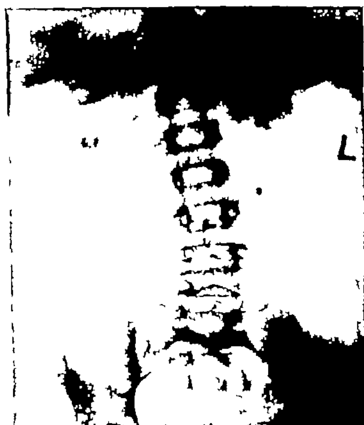
(*a*)

FIG 83—Renal calculus (left) with lumbar ureterectasis Girl aged 2 years 10 months (*a*) Skiagram showing calculus in left kidney, (*b*) excretion pyelogram confirming pelvic position of stone and demonstrating the ureteral dilatation

Referred by Dr Wyllie after two attacks of haematuria Urine heavily infected with *B proteus* There was no evidence of ureteric obstruction Pyelo-lithotomy was performed and the child was symptom-free for a year, when infection recurred and a small calculus was noted in the kidney The urine was sterilized by chemotherapy and there were no further symptoms for another two years The stone has recently been found at the lower end of the left ureter Urine sterile
(Composition of stone—phosphates)

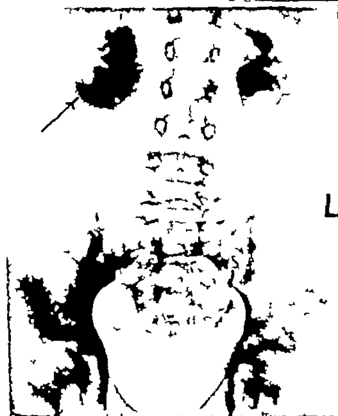


(*b*)



(*a*)

FIG 84—Renal calculus in congenital hydro-nephrosis. Girl aged 8 years. (*a*) Skiagram showing stone in right renal area, (*b*) excretion pyelogram demonstrating hydro-nephrosis (stone in lower calyx)



(*b*)

Child presented with severe attacks of pain in the right loin. Urine sterile. At operation an aberrant vessel was ligatured and divided, adhesions freed and the stone removed by pyelo-lithotomy. Three years later symptom-free, no recurrence of stone, and intravenous pyelogram showed only slight residual dilatation of pelvis.
(Composition of stone—phosphates.)

or the more specific symptoms of pain radiating down the penis and interruption of the urinary stream The mucosa is always inflamed and may be ulcerated, in late cases the bladder is severely contracted with hypertrophied muscle and inflammatory infiltration of the wall The diminished capacity is often associated with dilatation of the ureters and is, of course, responsible for severe frequency and incontinence Vesical calculi, if small enough, may be passed per urethram or may occasionally become impacted in the posterior urethra or at the meatus, causing acute retention.

Cystine stones

These are commonly multiple they may be renal, ureteral or vesical In the kidney the staghorn formation is common and there may be conglomerate stones formed by a fusion of a number of small ones The urine is sterile and acid in the first place, infection and dilatation, if they occur, are secondary to the calculus

The progress of the disease and its complications

Stones may, of course, be passed per urethram, but as in children they are often multiple and secondary to some other pathological process, the urinary tract is seldom able to clear itself by its own efforts The presence of stone inevitably leads to destruction of the renal parenchyma, rapidly where there is obstruction and hydro-nephrosis, slowly with fibrosis and atrophy where the urine is not dammed back The persistent infection causes recurrent fever, under-development, anaemia and wasting Where both kidneys are involved uraemia may supervene with all the signs described in Chapter 4

Children may sometimes, like adults, remain surprisingly well despite bilateral staghorn calculi, and a temptation to leave well (or comparatively well) alone may be very great, but a fatal termination before the child reaches adult life cannot be in doubt At times the child appears to have passed out of its stone-forming period, no more stones are formed where they might be expected, and after removal there may be no recurrence even where everything seems to favour it This fact is a considerable encouragement to undertake conservative surgery

Perinephric abscess may complicate renal calculi of long standing Its diagnosis and treatment do not differ from those in the adult and require no special description Calculous anuria we have not observed, though it has been reported in infants

Diagnosis

It must be emphasized that calculi in children can seldom be diagnosed from a history of typical renal colic, colic is rarely a feature of secondary infected stones, and even if it occurs the pain is not well localized in infancy or accurately indicated Diagnosis is reached either from the history of the passage of a stone or by the investigation of cases of pyuria, haematuria and recurrent urinary infection Certain points in this investigation, however, may be emphasized Vesical calculi and calculi impacted at the lower end of the ureter may be palpable with a finger in the rectum in small children, and this investigation should not be omitted Upon cystoscopy stones in the bladder are evident unless—which must be rare in children—they are in a diverticulum Cystoscopy may also give valuable information concerning the ureter the important points are discussed in Chapter 9 Most

stones are in fact diagnosed by radiography They may however be overlooked in skiagrams because of absolute translucency as in the case of pure uric acid stones, or relative translucency and obscuring of the field by gas and faeces in cystine or loosely formed phosphate stones Cystine itself is opaque to x ray but not very much more opaque than the surrounding tissues. Cystine stones, however often contain some calcium as well The opacities due to stone must be differentiated from calcification within the substance of the kidney due to nephro-calcinosis, tuberculosis and neoplasm These latter have a characteristic appearance and in any case the pyelogram is usually sufficient to make differentiation easy Pyelograms, of course are necessary to fix the exact position of the stones and to reveal any other pathology in the urinary tract.

The diagnosis should where possible include the cause of the stone not simply its presence This may require chemical examination as well as radiographic and particularly where there are multiple stones in sterile urine a metabolic cause should be sought

Treatment

Ideally treatment should consist of (a) the removal of the stone (b) the elimination of stasis in the urinary tract (c) the control of infection and (d) the elimination or minimization of stone forming substances excreted in the urine.

Unfortunately it is not always practicable to carry out this programme but it is as well to bear in mind that an operation of lithotomy is only the beginning of the treatment

Operations for renal calculi

In simple cases pyelo-lithotomy in children does not differ from the operation performed in adults, though as the kidney is somewhat lower it is often more easily delivered into the wound. In suitable cases pyelo-lithotomy can be combined with procedures for the correction of congenital hydro-nephrosis as described in Chapter 10 Dendritic calculi may sometimes be removed by pyelo-lithotomy provided that the pelvis is largely extrarenal and that the tips of the horns of the stone have not expanded within the calyces We do not hesitate however, to perform an extended nephro-lithotomy in order to remove a branched stone the function of the kidney after this operation is often surprisingly satisfactory The kidney having been delivered is held in the hollow of the assistant s hand his fingers compressing the pedicle, and an incision is made along the outer border which is deepened down to the pelvis and the stone picked out. It is helpful to have skiagrams taken of the exposed kidney during the operation in order to locate small secondary calculi hidden in the calyces in front of or behind the line of incision The bleeding is chiefly from small arteries lying close to the calyces, which can be picked up and ligated while the ooze from the parenchyma can be controlled by the mattress sutures which close the incision

The indications for nephrectomy in stone cases are very hard to define· no one doubts that a calculous pyonephrosis with no more than a shell of renal tissue and a normal contra lateral organ, should be removed, but with less severe lesions there is frequently difference of opinion Many surgeons feel that in removing a damaged, infected kidney they are removing a possible source of infection for the other kidney· our own inclination is to save the kidney wherever possible and rely

on the antibiotics to control the infection In recurrent dendritic calculi, where the other kidney is entirely normal, nephrectomy may, however, be justifiable even though the function of the affected kidney has remained good Where there has been obstruction at the pelvi-ureteric junction, and the dilated calyces remain as ideal sites for recurrence, a *partial nephrectomy, removing a wedge to include* the lower calyx, may be of assistance in obliterating an undrained area Hemi-nephrectomy may be practicable where the stone has formed in a double kidney, but too often the normal upper segment is too small to be worth the risk.

Neither Suby's Solution G nor ethylene diamine tetra-acetic acid have, in our hands, proved to be of great value, even where the kidney has been irrigated through a nephrostomy tube in the hope of dissolving fragments left behind at operation

Operations for ureteric calculi

The ureter can be approached in the child and the stones removed through the standard incisions At the same time, the elimination of stasis may require dilatation of the uretero-vesical junction, trans-vesical ureteric meatotomy, or on occasion the resection of redundant portions of the ureter In cases of megaureter, laying the dilated segment widely open to the bladder may not reduce the dilatation, but the flushing effect of the reflux is likely to prevent recurrence of the stone (*see* Appendix III, case 6) Where, however, there is a unilateral megaureter complicated by stone, a primary nephro-ureterectomy is often the wisest treatment.

Operations for vesical calculi

In our opinion litholapaxy has very little place in the treatment of children's stones in addition to the risks of trauma to the urethra it must be remembered that the stones are usually infected, that they may have formed around a foreign body, and that they may be composed of cystine, which substance is apt to jam the jaws of the instrument Supra-pubic lithotomy is easily performed, and in septic cases a short period of bladder drainage is all to the good.

Post-operative treatment

Operations for calculous disease must be followed up by medical treatment of the infection, and attempts to alter the composition of the urine in order to prevent recurrence The control of urinary infections is discussed in Chapter 5, but it may be mentioned here that mandelic acid derivatives, particularly Mandelamine, are of special value, since the acidification discourages the formation of phosphate stones The routine of an acid ash diet is difficult to maintain in childhood and we have come to rely on a high fluid intake and acidifying drugs The child's parents should be given apparatus for checking up on the acidity of the urine at regular intervals. It is neither practicable nor desirable to reduce the intake of calcium in the diet It has been customary to restrict the intake of fruit in patients with oxalate calculi, but it is very doubtful whether this measure is valuable Cystine stones are formed in an acid urine, and in these cases alkalis should be given to keep the *p*H at 8 or higher meat, which contains the sulphur carrying amino acids, should be restricted We are at present giving a trial to the practice, suggested by Shoer and Carter (1950), of administering aluminium hydroxide gels to reduce the urinary phosphorus It is anticipated that this measure will prevent or retard the recurrence of phosphate stones

CHAPTER 12

URINARY TUBERCULOSIS

ALTHOUGH the kidney is usually involved in miliary tuberculosis surgical disease of the kidney is not common in childhood indeed, pyuria of non tuberculous origin is so much more frequently encountered that there is some danger that the possibility of Koch s infection will be overlooked The disease in children does not however differ in any essential from the adult form and so long as it is borne in mind during the investigation of pyuria there should not be any exceptional difficulty in diagnosis

Pathology

Following the work of Medlar (1926) and Band (1935) it is now generally recognized that the earliest lesions of surgical tuberculosis are small multiple cortical foci in both kidneys At this stage there are no urinary symptoms but albumin and tubercle bacilli may occasionally be found in the urine and these lesions are the pathological basis of what was formerly described as tuberculous bacilluria* and are found in patients with other active foci in the body These early lesions are recoverable and in healing may leave very little trace of their presence a secondary lesion however may be formed in one of the renal pyramids (and this is usually a single lesion in one kidney) which undergoes caseation and finally ulcerates into the calyx. With the onset of ulceration pus cells and tubercle bacilli are constantly present in the urine, and the spread of the infection to the urinary passages may produce frequency haematuria and sometimes pain in the loin Tubercles in the mucosa of the pelvis may lead to obstruction at the pelvi ureteric junction with resultant hydro-nephrosis, and this obstruction facilitates the spread of the disease to other calyces Occasionally complete obliteration of the outlet converts the kidney into a pyonephrosis more often the disease assumes the ulcero-cavernous form and the renal substance is gradually excavated Some degree of calcification of the caseous focus can often be found from a very early stage Later on rings of calcification may outline each excavated calyx. The ureter is at first slightly dilated and atonic (*see* Fig. 85) but soon becomes thickened and fibrosed The bladder is at first affected only in the region of the ureteric orifice which is drawn upwards by the shortening of the fibrosed ureter tuberculous ulceration occurs in the vesical mucosa and an interstitial cystitis leads to severe contraction of the bladder The contra lateral ureter may become dilated either because of stricture at the orifice which is involved in the cystitis, or because of reflux from the contracted bladder Spread of the tuberculous

*We have had occasional instances under observation for several years. One girl, apparently otherwise in perfect health, passed undoubted tubercle bacilli in her urine over a period of years. She suffered no incontinence and led a normal life. The bacilli eventually disappeared from the urine and she remained well after being under continuous supervision for 8 years.

sease to the contra-lateral kidney may occur but it is more likely that a previously ent lesion has flared up Some authors have stressed the importance of a uberculo-toxic nephritis" which is said to affect the healthy kidney, the evidence r a specific nephritis of this type is not very impressive (Cibert, 1946) though nically it does appear that renal function improves after removal of the diseased gan



FIG 85 —Renal tuberculosis Boy aged 8 years Retrograde pyelogram showing lesion in left kidney

The child had a history of frequency for 6 months, and occasional haematuria An intravenous pyelogram showed a deformity of the upper calyx of the left kidney, associated with some stippled calcification, and slight dilatation of the pelvis and upper ureter At cystoscopy some inflammation of the trigone around the left ureteric orifice was noted The urine contained tubercle bacilli Three years after nephro-ureterectomy the boy continues well and symptom-free, and the urine sterile

linical picture

ince the tubercle bacillus reaches the kidney through the blood stream a primary ocus of disease must be present elsewhere in the body, though it may or may not e evident The earliest cases, and in children the most common (said to occur 17 per cent of all cases), are found among those undergoing sanatorium treatment for bone and joint tuberculosis Pus cells and tubercle bacilli may be found pon routine examination of the urine in symptomless cases, or the occurrence of aematuria and frequency maycall attention to the need for urological examination

In such children there are seldom any physical signs of the urinary disease and radiological examination may reveal nothing, or only the first signs of an ulcerated calyx. Where no extra renal focus is apparent, many cases are brought to light by the investigation of chronic pyuria, and more advanced disease with bladder involvement, is more likely to be found in this group Haematuria is a common form of onset, with a transient acute phase which may be mistaken for pyelitis Frequency perhaps with some burning pain on micturition is the rule though in the very young the onset of enuresis after a dry period may be the only evidence of this frequency In the later stages with a contracted bladder, supra pubic pain a continual urge to micturate and incontinence may reduce the child to a pitiable condition In older children aching pain in the loin may be a feature even in the early stages. The kidney is occasionally palpable though very great enlargement does not occur

In the majority of cases the disturbance to general health is not very considerable though tiredness apathy and loss of weight may be remarked upon The temperature is normal or shows only slight irregular rises The urine is clear only in the very earliest cases and a turbid acid urine without offensive smell is the characteristic finding. *B coli* and streptococci may be present as secondary invaders in the late stages Since the diagnosis ultimately rests with the pathologist, repeated examinations of the urine are essential with cultures or guinea pig innoculations if the examination is negative The first specimen of urine voided in the morning is the one most likely to contain the bacilli and is preferable to the usual 24-hour sample

Involvement of the male genitalia may occur in children as it does in adults and the diagnosis of tuberculous epididymitis should always indicate the need for a full urological examination In addition rectal examination should always be made in the urinary cases to detect thickening of the prostate or vesicles in the very young the thickened ureters may also be within reach of the finger

Pyelography

The extent of the disease can only be estimated from examination of pyelograms and though the intravenous series may be adequate, the very earliest change may only be evident in retrograde pictures There is indeed a certain danger of disseminating the disease by catheterizing the infected kidney and this procedure should only be undertaken after careful consideration Catheterization of the apparently healthy side will normally be undertaken to exclude bilateral disease.

The earliest pyelographic signs are fluffiness of the calyces with slight dilatation a filling defect or absence of one calyx due to spasm at the infundibulum Stippled calcification in relation to the affected calyx is of great importance in the diagnosis The whole pelvis and all the calyces may be somewhat dilated while one appears ragged. These early signs may be simulated by the slight hydro-nephrosis of chronic *B coli* pyelitis which may also produce spasm of some of the calyces, but the fluffy ragged appearance which may be seen only on a good skiagram and the calcification are diagnostic (*see* Fig 85) Some degree of ureteric dilatation is the rule In the later stages there may be well marked hydro-nephrosis with distortion of the normal outlines or the intravenous pyelogram may fail to show any evidence of secretion Diffuse calcification or ring shadows around the

excavated calcyces indicate advanced disease The bladder may appear contracted, and the opposite ureter and pelvis may be dilated without necessarily being involved by the disease Its calyces will then appear dilated but smooth.

Cystoscopy

The bladder may be entirely normal despite well marked deformity of the renal pelvis more often there is some oedema and inflammation in the neighbourhood of one ureteric orifice. Occasionally a few grey tubercles may be seen upon the mucosa with a more generalized cystitis, and later a definite ulceration The bladder capacity becomes considerably reduced and any attempts at distension may provoke bleeding from the ulcerated areas, which rapidly obscures the field The rigid drawn-up golf-hole appearance of the ureteric orifice is well known

Complications

Yates Bell (1949) has drawn attention to the fact that in renal tuberculosis associated with bone and joint disease, recumbency calculi may complicate the problem of treatment Epididymitis has already been mentioned as a possible feature in male children

Treatment

The place of chemotherapy (streptomycin, para-aminosalicylic acid, calciferol) in the treatment of urinary tuberculosis has yet to be defined No adequate series in children is available and we must at present be guided by adult experience (Jacobs and Borthwick, 1950)

In cases where tubercle bacilli are found in the urine, but in which there is no pyelographic deformity, treatment must necessarily be convervative, and will in fact almost certainly be directed to a focus elsewhere in the body Again, where the earliest lesions are found in a case undergoing treatment for bone or joint disease, it is often wise to defer urological treatment until orthopaedic procedures are completed In bilateral disease, where both kidneys are affected by advanced lesions, surgery is clearly inapplicable, and it is only with a sanatorium regime and chemotherapy that there can be any hope of improvement With these exceptions, however, the treatment of the disease must be primarily surgical, and there should be no question of "giving chemotherapy a trial" if the case is suitable for nephrectomy A course of streptomycin in conjunction with surgery, on the other hand, is undoubtedly of value (dosage of streptomycin 0 02 grammes per pound body-weight per day—continued for 3 months. P A S 1 gramme 3 times daily, concurrently)

The operation of choice is a total nephro-ureterectomy using two incisions, but there can be no doubt that this is a more serious operation than a simple nephrectomy, and may not be justifiable in a sick child The ureteric stump is apt to give rise to trouble, however, if left, and should tubercle bacilli persist in the urine after operation it is hard to know whether this indicates contra-lateral disease or simply residual infection in the stump

In many cases the vesical condition improves after the removal of the infected kidney, but if a severe contraction has already occurred the bladder cannot re-expand In such cases it is useless to give streptomycin even if there are actually tuberculous lesions in the bladder, as the normal effect of the drug is to increase

the fibrosis and a worsening of the frequency has been reported on several occasions If the contra lateral kidney is damaged by back pressure yet not infected uretero-colic anastomosis should be considered

Prognosis

Falci (1925) in an extensive review of published cases came to the conclusion that the prognosis in children was considerably worse than in adults Not only was bilaterality more common but the recurrence rate and mortality after nephrectomy was considerably greater Cibert and Perrin (1950) confirm the original impression that in children bilateral disease is more common and that the prognosis after nephrectomy is worse (in France)

Our own comparatively small series tends to confirm these views No doubt under sanatorium conditions many early cases do recover making the over all prognosis of the disease comparatively good In the more advanced cases, as seen by us, the outlook is gloomy Even when an apparently timely nephro ureterectomy has been done, the disease has proved lethal in a third of our cases

CHAPTER 13

NEOPLASTIC DISEASE

TUMOURS OF THE URETHRA

IN BOYS polyps in the posterior urethra springing from the verumontanum may cause persistent enuresis We have only met with a genuine polyp on two or three occasions

Fronds and tags of mucosa, possibly the early stage of true polyps, are sometimes seen

These lesions are for the most part readily destroyed by fulguration, but in some instances removal by cystotomy may be best

Winsbury-White (1948) has stressed the significance of such cases In our experience they are rare in young children

Sarcoma occurring in the urethra finds a rare mention in the literature. Joelson (1924) in a collected series of 24 included 2 children, a boy of 8 years and an infant Rhabdomyosarcoma (*see* later) may originate in the region of the prostate and any "urethral" sarcoma met with in childhood is likely to be of this nature

TUMOURS OF THE BLADDER

Haemangeioma*

Haemangeiomas are exceedingly rare in the urinary tract and very few have been recorded in childhood, though in some of the adult instances symptoms have been present since early life. It is perhaps remarkable that a lesion which is congenital should not declare itself more constantly at an earlier stage

Macalpine (1930) recorded 2 cases of vesical haemangeioma one in a boy aged 14 years, who had extensive naevoid formation in the scrotum, rectum and retroperitoneal tissues as well as in the bladder The boy was cured of his vesical bleeding by partial cystectomy Macalpine collected 19 other cases from the literature, 6 of which were in children He stresses the frequency of multiple and widespread haemangeiomatosis in the records

The following case was successfully dealt with by one of us many years ago at Great Ormond Street

D B , female aged 6 years

Presented with a history of 5 months severe haematuria with occasional passage of clots On cystoscopy a large ulcerating naevus of the fundus was seen with outlying patches The ureteric orifices were not involved (*see* Fig 86) Pyelography showed no abnormality of the upper tract Diathermy was carried out through the endoscope

*Haemangeiomas are to be regarded as congenital blood vessel anomalies rather than true neoplasms, but it is convenient to refer to them in this section The same applies to the lymphangeiomas

Three months later the bleeding having continued, the bladder was opened and extensive direct diathermy was applied Following this operation the bleeding ceased and a cystoscopy 3 months later showed a satisfactory scar During the subsequent years when she was under observation there was no further bleeding We believe that the child grew up normally

In 1944 one of us (T T H) was fortunate enough to be shown by Mr Leo Richardson a small boy under his care who had multiple wart like naevi in his bladder causing intermittent haematuria These were fulgurated by Mr Richardson but later resection was necessary

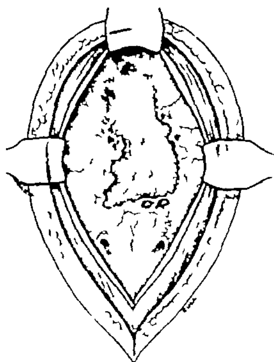


Fig 86 —Haemangeloma of bladder
Diagrammatic sketch at cystoscopy

Papilloma fibroma myoma

The villous papilloma of adult pathology has not been seen by us in the urinary tract in children We are not convinced that it ever occurs before adolescence Soft papillomas (sessile and pedunculated) do arise in the vesical mucosa as elsewhere in childhood Fibroma and myoma have also been described in the literature Any such tumour revealed by cystoscopy should certainly be adequately excised after cystotomy and its pathology established beyond doubt, since there is always a strong possibility of malignancy

Neurofibroma

Neurofibromatosis of the bladder is a possibility Kass (1932) described the case of a boy who had extensive cutaneous fibromatosis and developed haematuria On cystoscopy nodules were found projecting from a thickened bladder wall

In the following remarkable case in our series a tumour like formation at the base of the bladder in an infant girl caused acute retention and bilateral ureteral obstruction which proved rapidly fatal

W B , female, aged 1 month
Referred by Dr Wyllie The infant had much pain and difficulty in micturition The abdomen was very distended, and large tense rounded swellings were distinguishabl in both loins extending to the iliac fossae The bladder was distended and a solid mas was palpable apparently behind and below it, particularly prominent per rectum Blooc urea 69 mg per 100 ml A soft rubber catheter was passed with some difficulty a the urethra appeared to be deviated and tortuous 2 ounces of clear urine were with drawn with no appreciable change in the supra-pubic swelling The cystogram showec a small and apparently normal bladder

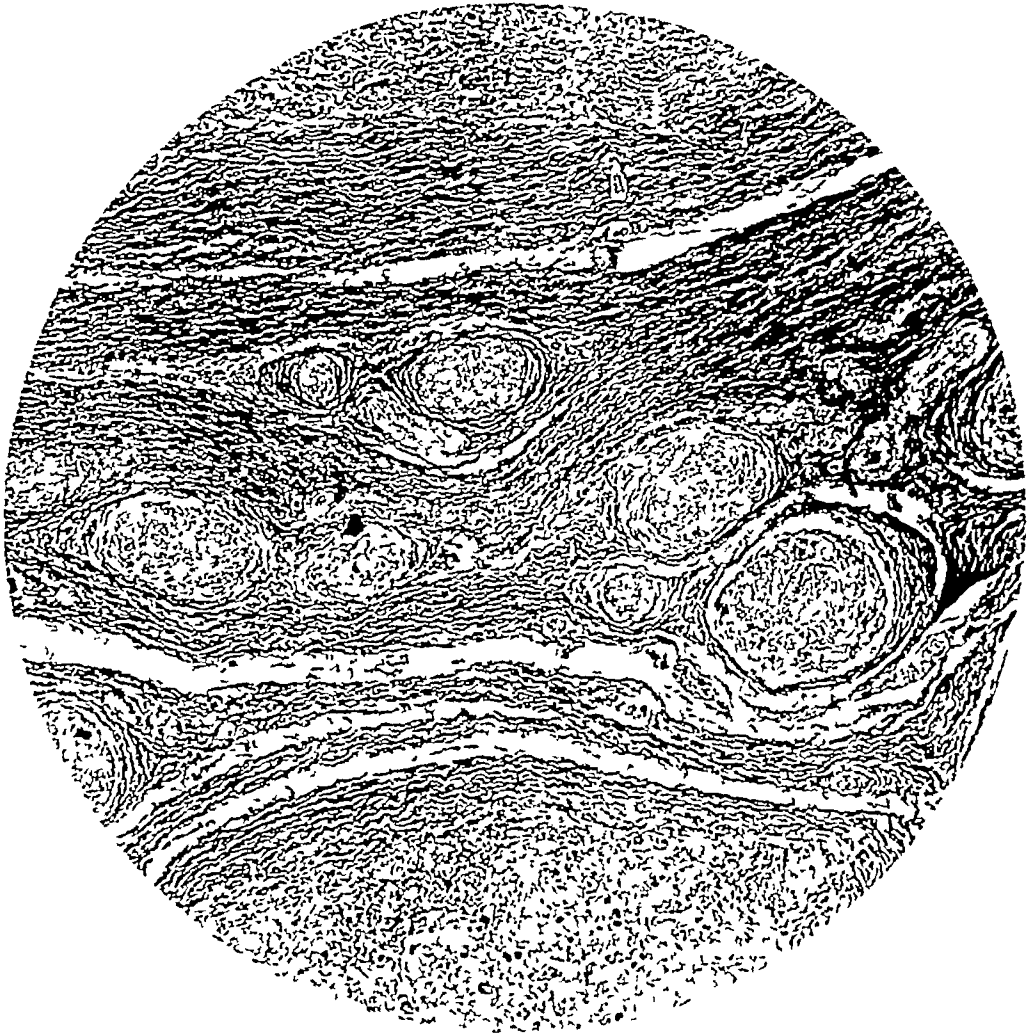


FIG 87 —Neurofibromatosis of bladder Photomicrograph (× 45) of section showing multiple nodular growths and many scattered ganglion cells in the vesical wall

Tentative diagnosis —A neoplasm with back-pressure distension
Operation —The right loin was explored and the kidney found grossly distended; opalescent, odourless urine was aspirated, gradually thickening to definite pus Nephrostomy was performed Following this procedure the blood urea came down to 26 mg per 100 ml and the infant's condition improved rapidly, natural micturition being resumed and the left kidney swelling subsiding The right nephrostomy continued to discharge urine Two months later there was a sudden setback with retention and dehydration, and the left kidney became tense again This was aspirated and 67

millilitres of clear odourless urine withdrawn The general condition remained unsatisfactory and the blood urea rose again Accordingly cystotomy was performed The bladder was found to be greatly thickened and the base bulged out by an extensive solid nodular mass, involving and obstructing both ureteric terminals and the internal meatus. A section was taken for histological examination The infant s condition remained too poor to permit of ureteral transplantation and in fact death ensued 7 days later

Pathology report —Death was due to retention back pressure and infection caused by the condition at the base of the bladder

The bladder wall was converted into a solid hard plaque like structure, 1–3 centimetres thick, with maximal involvement of the base. Thick muscle bundles could be appreciated within the otherwise homogeneous texture Both ureteric orifices and the urethral outlet were incorporated in the nodular mass Both ureters were dilated and thickened. The kidneys displayed intrarenal hydro-nephrosis, the left to a more marked extent, and there were small cysts and abscesses in the residual parenchyma.

Histologically the striking feature was the preponderance of well defined large neurofibromas with included ganglion cells in the stroma of hypertrophied smooth muscle and fibrous tissue. The arrangement of the tissues was that of a hamartoma rather than of a neoplasm (*see* Fig. 87)

Diagnosis —Neurofibromas arising from autonomic plexuses of bladder wall

Myxoma

Myxoma is usually mentioned as a benign tumour In the bladder apparent instances should be viewed with great suspicion as the following case illustrates

B D female, aged 4 weeks, was sent up to Great Ormond Street by her doctor who reported that at the infant s birth he had found a large cyst like tumour protruding from the urethra attached by a narrow stalk A ligature was tied round this and the tumour cut off The pathologist reported the growth as being a myxoma When the infant was cystoscoped a scar was seen on the trigone close to the left ureteric orifice Some vesicles and thickening were still present and diathermy was applied Three weeks later on further cystoscopy 1 or 2 residual vesicles were seen and the fulguration was repeated. At 1 year reinspection showed no sign of growth

At the age of 4 years the child returned on account of mild enuresis This had no relation to the original lesion and was soon rectified Opportunity was, however taken to examine the bladder again and the cystoscopic picture showed no abnormality

Unfortunately the specimen in this case was not available but by the helpful interest of Dr Kelly and Mr Barrington the slide was sent to us, of which Fig 88 is a photomicrograph The section is a typical rhabdomyosarcoma, confirmed by Dr Martin Bodian

A similar instance of a vesical myxoma attached to the trigone was found by Harris (1926) in a foetus He regarded it as a fusion defect between mesoderm (Wolffian duct) and endoderm (cloaca) entitling it a dysontogenic hamartoma

A myxoma will most often prove to be in reality a rhabdomyosarcoma as in the case here recorded which promises to be a fortunate instance of cure

Rhabdomyosarcoma (sarcoma botryoides)

This is the most important tumour of the lower urinary tract in childhood, though happily it is rare While we are dealing here with the growth as it originates in the bladder the same growth more commonly occurs in the vagina, the bladder being only involved later by invasion. Sarcoma botryoides has also been described in

the soft palate, and rhabdomyosarcoma without the grape-like formation, a somewhat different type of tumour, is, of course, met with in many situations

Sarcoma botryoides is a mesenchymal neoplasm of a most lethal character The histological components are embryonic mesenchymal cells capable of differentiating fully or partially into all varieties of adult mesenchymal tissues, that is muscle (striped and smooth), cartilage, and bone Yet it has its favourable features which would seem to offer some encouragement to radical surgery The growth, though rapid and locally invasive, is characterized by delayed metastasis, lymph gland involvement and blood stream invasion are late developments, and death

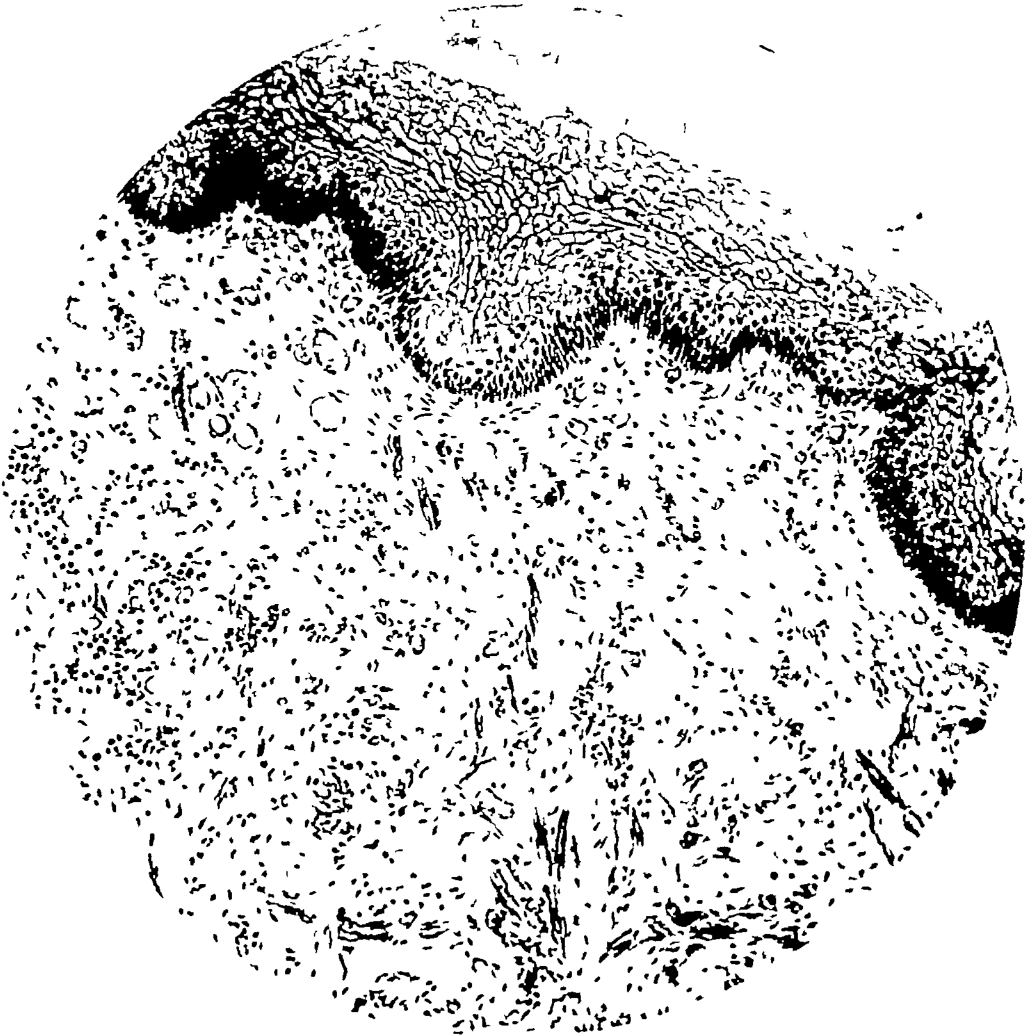


Fig 88 —Rhabdomyosarcoma of bladder Photomicrograph (× 90) of section showing neoplastic mass covered with transitional epithelium and consisting of loose embryonic mesenchyme of pseudomyxomatous appearance, and numerous striated muscle fibres

from the tumour *per se* may therefore only occur after several years Cappell and Montgomery (1937) described two instances in the palate (a) girl aged 10 years—died 7 years later with metastasis in lungs and lymph glands, and (b) girl aged 9 years—died 6 years later with metastasis in lungs and lymph glands Other

similar cases have been recorded by Nicory (1923) and Martin and Alexander (1924)

This delayed dissemination is in marked contrast, so far as children are concerned, to the renal embryoma.

A rhabdomyosarcoma in the bladder by reason of its local obstructive effects and the superadded infection should lead to early diagnosis On the other hand unless it be recognized early the rapid destruction of renal function will be fatal long before any metastasis of the tumour has occurred. Assuming that the growth is recognized before these secondary complications have supervened, there is a considerable hope that radical surgery provided it is radical enough, may be curative.

Pathology

The naked-eye appearances of these tumours are strikingly similar whatever the site of origin Arising from a tough solid base are seen masses of round, often polypoid and pedunculated, gelatinous nodules, which have been aptly likened to bunches of white grapes. hence the term sarcoma botryoides The grapes are often so loosely tethered by attenuated bands that they come away in the hand during manipulation

Histological examination reveals a pseudomyxomatous structure, which explains these remarkable appearances The cellular components of the tumours are relatively scanty predominantly embryonic, but differentiation into striated muscle fibres is commonly found and justifies the term rhabdomyosarcoma

When found in the bladder the tumour appears to originate in the area from the trigone to the bulb and the grape like clusters largely fill the bladder lumen

The traditional sarcoma of the prostate in boys of which there have been 4 cases in our series is a rhabdomyosarcoma The growth in these instances because of its confined environment seems very hard, almost rock like when felt bimanually at the bladder outlet The outflow of urine is seriously impeded and the bladder distended

The rhabdomyosarcoma grows rapidly and is locally invasive. It pushes its way into any neighbouring tube which happens to be available (the urethra or the ureter *see* case records below) But apart from this direct invasion and extension it does not metastasize by the blood or lymph stream until its later stages To repeat—in the case of the urinary tract its lethal nature which is devastating, is determined by its obstructive effects and the resulting infection rather than by its pathological potentialities As has been stated this makes it a rather more hopeful tumour than the renal embryoma if diagnosed early and treated radically

Clinical features

The earliest manifestations are those of persisting infection with marked disturbance of micturition (frequency urgency and unexpected incontinence and dribbling) We have been impressed in our cases by the degree of pain, spasmodic and severe, making the child scream and strain This is probably explained by partial prolapse into the urethra of a grape like appendage

Sooner or later a little bright red blood will be passed with the urine, usually with a bout of straining, and the bladder may well be distended Unfortunately the significance of the incipient disturbance of micturition is seldom appreciated

and the child is usually, when first seen, gravely ill with a distended bladder and enlarged tense kidneys In such circumstances, on passing a catheter perhaps 3–4 ounces of blood-stained urine are withdrawn without much appreciable diminution in the size of the bladder. Further palpation now enables recognition of the solid tumour filling the bladder and, bimanually with a finger in the rectum, its consistency and outline can be further appreciated

Cystography will demonstrate the tumour outline In the early case a cystogram may be of inestimable value in leading to diagnosis (*see* Fig 89 (*a*) and (*b*) and record of case C H) Cystoscopy enables confirmation of the nature of the lesion by inspection

Treatment

Radiotherapy

The rhabdomyosarcoma is not "radio-sensitive", but as Cade (1951) has emphasized, "radio-resistance" should not be taken to mean that the tumour cells are uninfluenced by the radiation, or that radiotherapy has no place in the management of radio-resistant growths Appropriately applied it may profoundly modify the rate of division, and indeed metamorphose the nature of the component cells of the tumour and so reduce the rapidity and invasive characters of its progress.

Pre-operative radiotherapy, by reducing the inherently malignant nature of the component cells of such tumours in this way, may be expected to ensure greater safety and success to their surgical removal, though the size of the tumour may not be materially reduced Delay in operation may be a wise insurance premium to pay In the growth now under consideration, however, any possible advantage from pre-operative radiotherapy has to be carefully balanced against the obvious disadvantage of increasing deterioration of renal function It is of no advantage to the child to "soften up" the growth by preliminary bombardment, if renal function is in the meantime being seriously jeopardized

Operative treatment

It must again be emphasized that a wide operative removal offers the only hope of cure, and no time should be wasted in needless investigation Pre-operative radiotherapy will rarely be feasible in the bladder growths

For practical purposes any solid tumour of small proportion discovered in the bladder of a child by a fortunate cystoscopy should be regarded as potentially malignant and adequately excised after cystotomy Unfortunately, however, the rhabdomyosarcoma will rarely be caught in so favourable a guise We may expect to be faced with a very large tumour so seriously implicating the bladder that nothing short of a total cystectomy will achieve its complete extirpation Obviously this is a formidable procedure in a small child Nevertheless we have carried it out in two cases without undue anxiety

The vital consideration when the child is first seen is the condition of the upper urinary tract and the renal function With distended "back-pressure" kidneys and seriously embarrassed renal function, a bilateral emergency nephrostomy must be done at once Combined with this, appropriate intravenous transfusion and chemotherapy must be instituted in order to resuscitate the child When the general condition has been sufficiently restored, the further treatment

89 —Case C.H Rhabdomyosarcoma of
ıdder
) Cystogram showing filling defect in bladder and bilateral reflux
) Operation specimen (total cystectomy) Bladder in section. Note extension of growth into urethra. Terminal urethra subsequently excised.
ıild alive and well 3 years later



(a)

(b)

can be planned In happier circumstances (*see* case C H , later) preliminary nephrostomy may not be necessary, and the major operation can be proceeded with at once

Stage 1—The ureters are transplanted to the sigmoid, one at a time, with a fortnight's interval (*see* page 230), the ureters being divided as far above the bladder as seems feasible

Stage 2—As soon after this as possible (2–4 weeks according to the child's condition) the final removal is undertaken, with every possible aid from blood transfusion and so on

Cystectomy

The technique of the operation as outlined by Millin and Masina (1949) is applicable to the child, and, from our experience, is somewhat easier to perform in the young owing to the greater accessibility of the bladder The utmost gentleness is essential, particularly to avoid any rupture and spilling of the growth The real difficulty is to ensure wide enough removal of the urethral aspect In the girl, it will probably be easier to remove the urethra entire in the boy, the problem is more difficult In our case T.C (*see* Fig 90) it was found possible to get well below the growth without undue difficulty Where the prostatic urethra is seriously involved, the approach would have to be extended by division of the pubic symphysis.

CASE RECORDS

1 C H , female, aged 2 years 3 months

Healthy and well till 18 months of age Then had attack of "cystitis and haematuria". Was treated with sulphonamides "successfully" General health deteriorated with symptoms of increasing frequency incontinence and polyuria No further haematuria, but marked thirst, loss of weight and abdominal pain

On admission to Great Ormond Street, under the care of Dr Wyllie, the child was very ill, wasted, anaemic and dehydrated The abdomen was distended and both kidneys palpable A centrally placed tumour, the size of a tennis ball, was felt in the pelvis, clearly the bladder Urine thick, purulent, (*B coli*) Residual urine 2 ounces Blood urea 83 mg per 100 ml , Hb 42 per cent , R B Cs 2,900,000, W B Cs 14,700 Cystogram showed reflux bilateral megaureter and hydro-nephrosis, and a filling defect at the base of the bladder (Fig 89 (*a*)) Continuous catheter drainage of bladder and intravenous fluids improved her condition Blood urea fell to 47 mg per 100 ml Cystoscopy now showed a large polypoid growth filling the bladder, apparently springing from the base

Operation—Cystotomy Bladder greatly hypertrophied and filled with growth—typical sarcoma botryoides The growth apparently sprang from the lower half of the anterior wall The prominent portion was moved by diathermy for histological examination, and the diagnosis of rhabdomyosarcoma confirmed One month later the right ureter was transplanted into the sigmoid, and the left ureter 12 days later Following the transplantation the child's general condition improved greatly, though the blood urea rose to 101 mg per 100 ml

After 5 weeks interval total cystectomy was performed The tumour was apparently completely enclosed by the bladder wall and the operation uncomplicated (*see* Fig 89 (*b*)) There was some uncertainty about extension into the urethra at the level of section, and the urethral remnant was finally removed from below before her discharge

Approximately 3 months later the child went home in very good condition Rectal control was promising, dry by day with occasional wetting only by night Blood urea 76

mg. per 100 ml Complete control by day and night was gradually established, and 2 years later she was symptom free and in excellent health and has remained so to date (3 years)

Dr Bodian s report is as follows.

Naked eye examination—The specimen consisted of the tumour-filled and somewhat contracted urinary bladder the 2 ureteric stumps and the posterior end of urethra A piece of skin containing the cystotomy wound was attached to the anterior bladder wall The maximum diameters of the bladder were 7 5×4 2×4 6 centimetres The weight of the specimen was 100 grammes. The length of the posterior end of the urethra measured 1 3 centimetres It was distended with growth. Both ureteric stumps apparently contained growth the right more than the left Also received was a lymph node of 1 2×1×0·7 centimetres size, which was greyish white and firm. It had been removed from the left pelvic fossa.

A mid sagittal section was made through the bladder The lumen of the viscus was found to contain a great deal of grape-like pseudomyxomatous yellowish white growth which occupied all the lumen except the uppermost fundus. The bladder wall was thickened and trabeculated The growth apparently arose from both anterior and posterior walls of the lower half of the bladder (below the site of cystotomy) and from the walls of the posterior end of the urethra The thickness of the anterior wall was increased up to 1 2 centimetres, the posterior to 0·9 centimetre (*see* Fig. 89 (*b*))

Microscopy—The lymph node from the left pelvic region displayed no histological evidence of neoplastic change

The new growth appeared to arise from the submucous coat of the distal half of the anterior bladder wall and extended down to the cut edge of the urethra. The neoplasm displayed mainly the characteristics of an undifferentiated mesenchymal sarcoma (fibrosarcoma) but scattered areas contained groups of muscle fibres with cross or longitudinal striation

One or two areas showed mucinous interstitial stroma The muscle coats of the bladder walls were not invaded by the new growth, except for 1 or 2 very small areas.

2. T C. male, aged 1 year 9 months

Three months history of incontinence and dysuria. Abdominal pain and recent haematuria. Vomiting during week preceding admission On admission fractious and very miserable well developed child, good colour puffy under eyes, heart and lungs normal. Bladder apparently distended up to umbilicus. A rubber catheter passed easily and 3½ ounces clear urine were withdrawn The supra-pubic lump remained much as before. On compressing this a small quantity of very blood-stained urine escaped from the catheter which was then withdrawn Further examination of the abdomen confirmed the presence of a rather hard lump resembling a distended bladder smooth, symmetrical and freely mobile from side to side. Bimanually with a finger in the rectum, this was clearly a solid tumour of the bladder Both kidneys could be felt enlarged and tense. Blood urea 346 mg. per 100 ml W B Cs 11 500 (73 per cent poly., 26 per cent lymph. 1 per cent mon.) Tentative diagnosis tumour or cyst of bladder leading to obstruction and back pressure Cystography failed (catheter obstructed)

An immediate bilateral nephrostomy was performed. Both kidneys tensely distended. Following operation 40 ounces urine drained in the first 12 hours (urea content 1 per cent) On the day following operation blood urea fell to 155 mg. per 100 ml but urine output from kidneys dropped rapidly (2nd 12 hours after operation 9 ounces only) Put on to intravenous drip (glucose 5 per cent in water 3 pints in 24 hours followed by mixture 3 pints glucose (5 per cent) water 1 pint glucose (5 per cent) N/2 saline) Condition steadily improved. Puffiness of eyes disappeared. The nephrostomy tubes were removed after 6 days Blood urea now 50 mg. per 100 ml. BP 120/60

At this stage it was decided that the diagnosis was almost certainly a malignant neoplasm, and that radiotherapy should be started in the hope that shrinkage might diminish the ureteral obstruction preliminary to a laparotomy Following the first exposure the child was not so well Temperature rose to 103° F The treatment was stopped He now passed through some very anxious weeks, during which there was obviously a severe infection The nephrostomy wounds discharged actual pus at times and the blood urea rose to 95 mg per 100 ml With the aid of penicillin, sulphacetamide and appropriate transfusions, under the unremitting care of Dr Hugh Jolly, he survived and improved



FIG 90—Case T C Rhabdomyosarcoma of bladder Operation specimen (total cystectomy) Bladder in section Death 9 months later from extension of growth to right kidney and local recurrence in the pelvis

Seven weeks after his admission cystotomy was performed *This confirmed* the presence of a rhabdomyosarcoma, apparently springing from the trigone but completely filling the bladder The appearance was typical—a firm red fleshy solid infiltrating tumour with masses of grape-like "cysts" on the surface (*see* Fig 90) (Dr Bodian's report on the specimen is appended)

Following the cystotomy the child's general condition improved; drainage and lavage of the bladder were carried out Three weeks later it was decided to transplant the ureters and if possible attempt total cystectomy The right ureter was transplanted into the sigmoid (ureter found grossly thickened and apparently involved by growth) Two weeks later left ureter transplanted (on this side the ureter was normal) After a further 2 weeks blood urea 82 mg per 100 ml Condition good Total cystectomy carried out

After a further month at a convalescent home his blood urea was 85 mg. per 100 ml but general condition very good.

Three months later pains in abdomen large swelling in left iliac fossa felt per rectum left leg larger than right no pitting right kidney palpable. One week later right kidney full—very large left kidney full Mass in left iliac fossa larger and more fixed Right nephrostomy scar incised and pus let out. On pressure gelatinous material resembling the original growth was obtained and histology confirmed this Eight weeks later he died

Histological diagnosis—Rhabdomyosarcoma of bladder prostate and seminal vesicles

Pathological report (operation specimen)

Naked eye examination—The specimen weighed 204 grammes. It consisted of bladder prostatic gland and neoplastic growth

External examination—This revealed a roughly L shaped structure The upright limb measured 9 centimetres in length 5 5 centimetres in maximum lateral diameter and 5 5 centimetres in maximum antero-posterior diameter The horizontal limb measured 7 5 centimetres in length, 4 centimetres maximal height, and 5 5 centimetres in maximum lateral diameter The anterior upright limb was formed of bladder distended by tumour which had protruded through the ruptured fundus The fungating growth extended beyond the aperture of the bladder for up to 5 centimetres and was formed of large bunches of grape-like growth which was partly very firm, and in other parts more elastic, with haemorrhage

A mid-sagittal section was made

The left half displayed a thickened posterior bladder wall of up to 8 millimetres thickness, from the lower part of which arose a fungating greyish white growth along a 2 centimetres wide base.

The anterior bladder wall showed diffuse infiltration with whitish growth, and its thickness measured up to 2 centimetres. A large fungating mass arose from the antero-lateral portion of the fundus. Another very large fungating mass arose from the base of the bladder extending upwards into the lumen on a long stalk The lumen of the bladder was almost occluded by growth.

A markedly enlarged prostatic gland was attached to the base of the bladder Its wall was very thick The outer part appeared normal grey prostatic tissue, and the inner part was formed of white tumour tissue Its lumen communicated with the distended prostatic urethra and several small neoplastic grapes protruded from the prostate into the prostatic urethra beyond its cut end

Both vasa deferentia and vesicles were attached to the left half of the specimen It appeared that the left vesicle was considerably enlarged with growth

The left ureter could be discerned a probe could be passed in to an extent of 1 2 centimetres The extravesical length of the ureteric stump was 4 millimetres.

The right half of the specimen showed almost identical appearances with the exception of the following features

(1) The supra-pubic cystotomy was attached to the anterior wall

(2) The right ureteric stump was distended with growth. The uretero-vesical junction measured 2×1 25 centimetres in maximum diameters. A knob of growth protruded a little beyond the cut section of the ureter which measured 0·6 centimetre in diameter The tumour totally blocked the lumen of the ureter

(3) A large neoplastic grape (2×1 5×1 5 centimetres) protruded beyond the cut section of the prostatic urethra and blocked its lumen completely

Microscopy—Representative blocks were examined from anterior and posterior walls of the bladder bladder base, prostate, seminal vesicles, and the growth in the bladder lumen.

The previously made diagnosis of rhabdomyosarcoma was confirmed It appeared to arise from the lower half of the anterior bladder wall, the bladder base and the prostate, and to infiltrate the anterior bladder wall to the fundus The posterior wall was *not infiltrated, but growth in the lumen was closely applied to it* Both seminal vesicles were invaded by growth

The growth in the lumen was largely of a spindle cell type, the growth in the bladder and prostate walls largely formed of myoblasts, about 10 per cent of which displayed some evidence of cross striation

TUMOURS OF THE KIDNEY

The outstanding renal tumour of childhood is unfortunately the deadly embryoma, but innocent growths are not unknown

Fig 91 shows a lymphangeioma which occurred in a girl of 4 months The infant presented with a large mobile renal tumour, which was removed by Sir Lancelot Barrington-Ward in the understandable belief that it was an embryoma

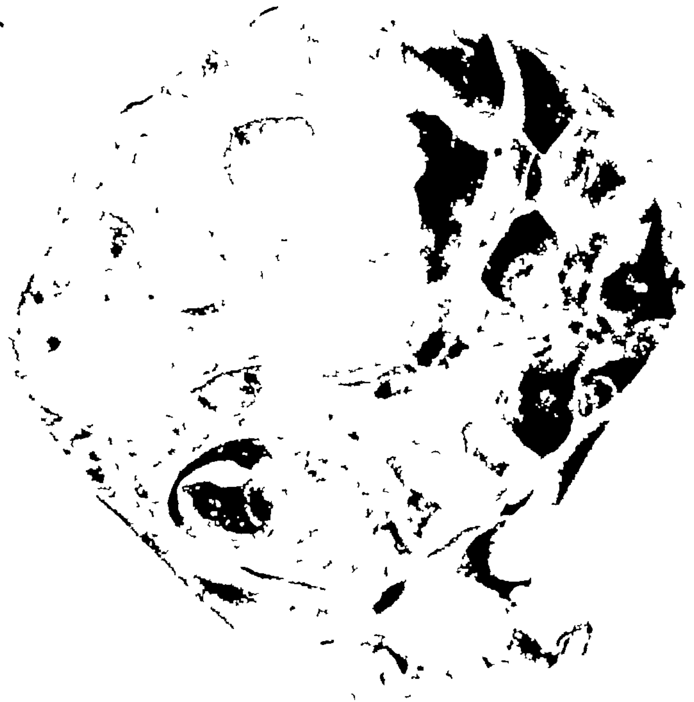


FIG 91 —Renal lymphangeioma Sir Lancelot Barrington-Ward s case Museum, Hospital for Sick Children.

(By courtesy of the Journal of the Faculty of Radiologists)

Mr Denis Browne had the good fortune to meet with a similar case After the nephrectomy his patient was found to have a tumour in the chest. This was successfully removed and proved to be a lymphangeioma also—apparently of independent origin

Renal haemangeiomas are an exceedingly rare possibility in childhood. There has been no case in our series Swan and Balme (1935) reported a case in a 15-year

old schoolboy in whom nephrectomy was necessary for severe recurring haema turia

As previously stated the villous papilloma of adults is virtually unknown in the urinary tract of young children One case of papilliferous carcinoma of the renal pelvis occurred in an infant girl of 18 months many years ago at Great Ormond Street. Nephrectomy was performed by the late Mr George Waugh but the child died some months later from metastases (Sections of this growth have been confirmed as carcinoma by Dr Bodian)

Secondary renal deposits are seen in the kidney for instance from lympho-sarcoma elsewhere, and neuroblastoma

Nephroblastoma (embryoma, Wilms tumour)

This is traditionally styled Wilms tumour since Wilms classical paper in 1899

Pathology

The tumour is clearly of embryonic origin and is therefore most correctly designated a nephroblastoma As to whether it derives from proliferation of undifferentiated cells in the Wolffian ridge (Wilms 1899) or from cells of the metanephros (Busse, 1899) can only be a matter of speculation without practical significance The tumour contains mesenchymal and epithelial elements exhibiting every variety of differentiation and disorderly arrangement. Histologically we see fibrous tissues, striated and smooth muscle cartilage and bone together with primitive glomeruli and renal tubules The tumour tends to outgrow its blood supply and areas of necrosis are common as well as haemorrhage from rupture of large immature blood vessels Occasionally calcification is seen

The proportion of the various cell elements and the degree of their differentia tion vary remarkably and in consequence individual tumours differ widely in general configuration and histological structure while different areas of the same tumour afford quite distinctive appearances

A glance at Figs 92–98 (pages 196–199) showing the cut-surface appearances of a series of tumours, and Figs 103–107 (pages 203–207) histological photo-micrographs will serve to emphasize to the reader this striking variability of structure Very often the growth is demarcated by a well defined pseudocapsule, which in reality consists of compressed atrophic renal tissue sometimes partially replaced by fibrous tissue Soft areas of haemorrhage and necrosis are common Fig. 92 shows a small growth in the substance of the kidney which had filled the pelvis with blood and led to severe haematuria. Some tumours show many cyst like spaces containing grumous or blood stained material and occasionally papilliferous growth (*see* Fig. 93)

The practical point arises as to whether this variability of structure can be construed as an index of relative malignancy whether we can say from a detailed histological analysis, that this or that particular growth is more malignant or less so Unfortunately we have not been able as yet to establish by clinico-pathological correlation any helpful guide to prognosis along these lines

It is however clear to us that the histological structure of the individual tumour is a reliable index of its radio-sensitivity the higher the proportion of fibro-muscular non-epithelial elements and the more highly differentiated the component cells, the more radio-resistant the growth and *vice versa* and to a



Fig 92

FIG 92—Nephroblastoma Small growth with haemorrhage into renal pelvis and ureter Severe haematuria led to early nephrectomy in this case but death ensued 9 months later from lung metastasis

(*By courtesy of the Journal of the Faculty of Radiologists*)



Fig 93

FIG 93—Nephroblastoma (L) removed from baby boy aged 1 year 7 months, showing total destruction of renal tissue and remarkable degree of differentiation with cystic formation

This child presented with a massive symptomless tumour in the left loin which did not transilluminate Excretion pyelography revealed a normal right kidney the left being completely silent Pre-operative irradiation produced no shrinkage of the tumour Following nephrectomy he was given a further course of irradiation He remained well for 3 years and 3 months when death rapidly ensued upon a local recurrence

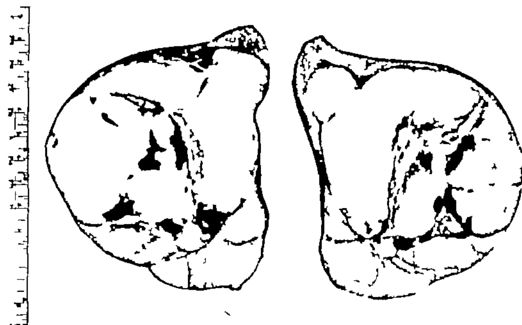


Fig 94 —Nephroblastoma, removed from an infant boy aged 7 months (whole specimen weighed 450 grammes) No irradiation. Alive and well (4½ years)

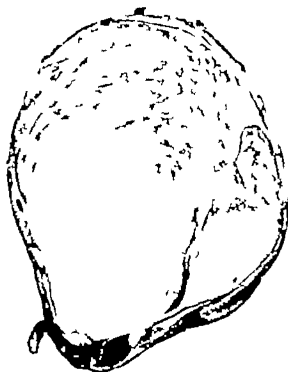


Fig 95 —Nephroblastoma Tumour removed from infant boy aged 1 year 6 months. Histology showed a high degree of differentiation in the cellular element. Post-operative irradiation only Child well so far (2 years)



Fig 96

FIG 96—Nephroblastoma Tumour removed from a boy aged 2 years Histologically the tumour elements showed a remarkable degree of differentiation into smooth muscle fibres. No pre-operative irradiation but post-operative course given So far alive and well (2 years 9 months)



Fig 97

FIG 97—Nephroblastoma Small tumour in upper half of bifid kidney (left)

(By courtesy of the Journal of the Faculty of Radiologists)

Girl aged 2 years Presented with haematuria and infected urine Excretion pyelography showed bilateral duplication of upper tract, the upper half of the left kidney being silent Growth found at operation nephrectomy performed A course of irradiation was subsequently given but death ensued 6 months later from lung metastasis



Fig. 98

Fig. 98.—Nephroblastoma. Complex tumour recently removed showing well marked pseudocapsule.



Fig. 99

Fig. 99.—Nephroblastoma in a fused kidney. Retrograde pyelogram from a boy aged 5 years, who presented with a large rounded medial firm abdominal tumour reminiscent of a fibroid. An exploratory laparotomy was performed but the growth proved to be inoperable. Death ensued 9 months later.

certain extent it is possible to take a slightly more hopeful view of this radio-resistant type of tumour We also see why a proportion of the tumours, as seen clinically, fail to show any dramatic shrinkage in response to radiotherapy

Relation of the tumour to the kidney

This also is variable The growth may be so intimately intrarenal that the kidney tissue is reduced to a thin peripheral shell, and may even be no longer detectable (*see* Fig 93) On the other hand, in not a few cases the neoplasm is almost entirely extrarenal

Fig 97 shows an embryoma occurring in the upper half of a double kidney, this caused haematuria which led to early diagnosis by pyelography Fig 99 is a pyelogram demonstrating a growth in the middle of a horse-shoe kidney

Bilateral tumours are on record We have not recently met with an instance

Effects on the renal pelvis

The effects of the growth on the pelvis are important from the point of view of diagnostic pyelography When the renal tissue and pelvis have been totally obliterated, the pyelogram will draw a complete blank on the affected side It may be noted that in a hydro-nephrosis of dimensions likely to be confused with an embryoma, the pyelogram will be equally silent

In the majority of instances, however, some portion of the kidney remains actively secreting and some fragment of pelvis available The pyelographic appearance most often seen is of a distorted segment of the pelvic shadow situated anywhere in the tumour area There may, however, be a normal pelvic segment with gross distortion of the remainder

Dilatation of the pelvis is comparatively rare, but we have seen it on a few occasions (*see* Fig 100) Generally speaking, if the pyelogram shows a hydro-nephrosis of any size, doubt is thereby thrown on the diagnosis of embryoma

Mode of spread

The renal embryoma grows with alarming rapidity It is at first reasonably defined, though it soon acquires fixation to surrounding structures by peripheral adhesions. It is thus common to find it necessary at operation to excise adherent peritoneum The adrenal is usually incorporated and removed with the tumour mass Separation from the diaphragm and lumbar muscles seldom presents difficulty, but in the later stages direct invasion of these may occur, probably through the medium of rupture of softened necrotic areas and escape of tumour cells Tanner (1943) recorded a case of intraperitoneal haemorrhage from such a rupture

The growth will push its way into any available tube In one of our cases (which still survives after 17 years) the upper ureter was filled with non-adherent growth

The lymph glands in the vicinity of the renal pedicle and vessel roots are usually enlarged and should be carefully peeled away as they may be implicated

The sinister characteristic, however, is the ready spread to the veins The venous components of the tumour are very large ill-formed blood spaces, which are readily invaded, and the way to the renal vein is wide open (Fig 101 shows a remarkable instance in which the growth can be seen extending up the vena cava



Fig. 100

Fig. 100.—Nephroblastoma Excretion pyelogram showing distortion and some dilatation of pelvis (right kidney)

Girl aged 4 years. The tumour in this case was very large and extended far down into the right iliac fossa and across the mid-line. When first seen it was regarded as inoperable. Irradiation reduced the size so considerably that the growth was ultimately removed. Despite a further post-operative course of irradiation, however, death ensued 4 months later.

Fig. 101.—Specimen from the Museum, the Hospital for Sick Children, showing a left nephroblastoma with direct extension of growth up the vena cava into the right auricle. This boy was admitted with severe haematuria and was too ill for any operation to be attempted



Fig. 101

and actually filling the right auricle) Intravenous embolic metastasis is the surgeon's nightmare in these grim cases

Lung metastases are the most common form of haematogenous dissemination and are usually the immediate cause of death, but secondary growths may be seen on occasion in the liver, peritoneum and elsewhere Subcutaneous deposits occurred in two of our cases, one in the groin and the other in the scalp

Clinical features

Most of these growths are seen in the first 3 years of life, many in infants under 12 months, and not a few under 6 months The onset is insidious and there is usually no hint of anything wrong until a prominence is noticed in the child's abdomen In some cases, haematuria may occur and lead to early recognition (*see* Figs 92 and 97) We have known it to follow palpation of the lump Unfortunately it is not a common sign and the urine in most cases is perfectly



FIG 102 —Kidney (left) removed from a girl aged 4½ years showing the effect of irradiation on her nephroblastoma This child presented with a huge abdominal tumour which shrank with dramatic rapidity under irradiation to the size shown in this operation specimen Four months later the growth had recurred locally and grown to a size comparable with the original tumour Despite further irradiation she died 6 months after the nephrectomy

(By courtesy of the Journal of the Faculty of Radiologists)

normal When the child is first seen, the abdominal swelling is often very obvious to (trained) inspection, the tumour mass is very large and the growth already far advanced

In such circumstances when an embryoma is suspected, palpation of the lump should be as gentle and brief as possible It is essential, of course, to determine

e character and extent of the swelling, but this should be done without any edless manipulation An embryoma should never be pawed about for teaching irposes. this is very likely to sign the child s death warrant The essential features ving been determined by one observer no one else should repeat the palpation

iaracteristics of the lump

ie typical tumour is felt as a firm rounded solid lump in the loin thrusting rwards and extending inwards to the mid line or beyond The surface is often bulated and softer areas may be distinguishable The tumour mass is dull to

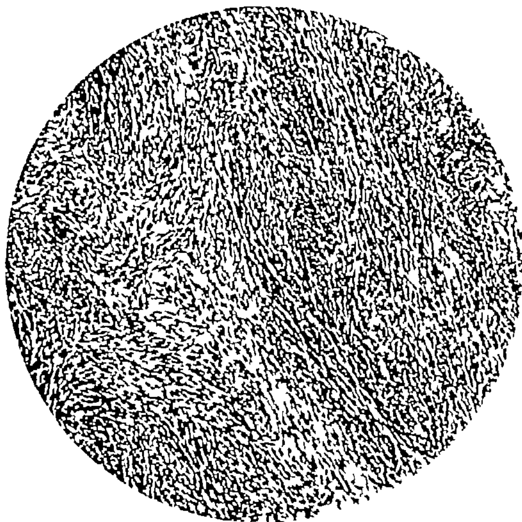


FIG 103 —Photomicrograph (× 90) of section showing differentiated smooth muscle and fibrous tissue.

By courtesy of the Journal of the Faculty of Radiologists.

ercussion though colon resonance may be recognizable on the anterior aspect. t does not transilluminate

Individual tumours may differ in their siting. An upper pole tumour will ormally be at a higher level and may bear some resemblance to an enlarged pleen Lower pole tumours tend towards the iliac fossa

Such a hydro-nephrosis will give a completely silent pyelogram, and this, though not unknown, is rare in embryoma (*see* above) In such a case cystoscopy may permit successful decompression through a ureteric catheter and so provide final confirmation

Neuroblastoma—Distinction may be difficult but is usually possible The neuroblastoma is generally more deep-seated and medial in position, and more fixed and nodular in character Outlying nodules in the line of the sympathetic chain are sometimes a feature (suggesting glands) The pyelogram is often of the greatest

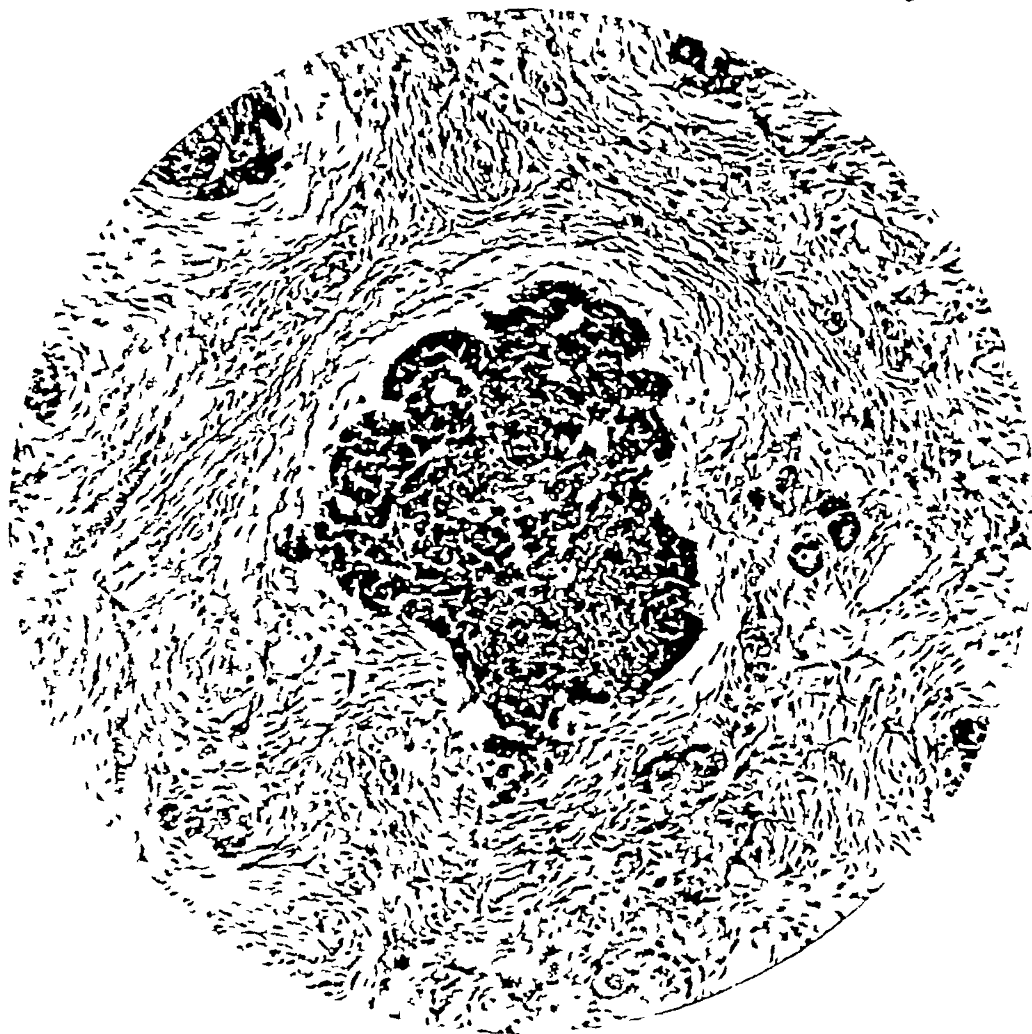


FIG 106—Photomicrograph (× 100) of section showing mesenchymal tissue with cluster of epithelium
(By courtesy of the Journal of the Faculty of Radiologists)

value in a doubtful case In the neuroblastoma the outline of pelvis and calyces is usually complete, but the whole pyelogram is displaced rather than seriously distorted (*see* Fig 109) Calcified areas are seen in these tumours more frequently than in the nephroblastoma, but the point is by no means diagnostic (*see* Figs 109 and 111)

Pararenal teratoma—This exceedingly rare growth must be mentioned as occurring in infancy One instance was met with in an infant at Great Ormond Street and

the growth successfully removed Its nature was suspected before operation from the bony shadows in the skiagram Occasional cases have been recorded by others (for example Campbell 1933)

Treatment of the embryoma

In the light of present knowledge the compelling indications are operation at the earliest possible moment it can safely be undertaken and ligation of the renal vein as early and expeditiously as possible during the operation It is the writers conviction that failure to keep these two over riding principles foremost in mind



FIG 107—Photomicrograph (× 90) of section showing development of cystic spaces. (*By courtesy of the Journal of the Faculty of Radiologists.*)

accounts for at least a proportion of the disappointing results in the cases in our series Our experience has convinced us that routine pre-operative therapy is a mistake

Decision as to immediate operation

Some tumours when seen are so massive that immediate operation clearly implies a formidable hazard and considerable manipulation before the renal vein can be

isolated If soft areas are clinically detectable on the tumour surface, the danger of rupture during operation is a very real one It is in such circumstances that pre-operative radiotherapy is still employed by us One may anticipate rapid shrinkage and firming up of the tumour In many cases the shrinkage is quite dramatic (*see* Fig 102, page 202) Unfortunately it is the softer, rapidly growing and "undifferentiated" tumours which respond so strikingly, and these appear to be the most highly malignant Very rapid shrinkage is, in our experience, of ill omen.

Following the preliminary irradiation, there is an optimum moment for operation, namely within 7–10 days of the final dose After this period the reactionary adhesions become progressively firmer and separation of the tumour is likely to prove correspondingly more difficult, tiresome and dangerous

Operation

Preliminaries

A "drip" transfusion is usually started a few hours before operation, and blood should be given if the haemoglobin percentage is unduly low The transfusion is maintained through the immediate post-operative period as may seem necessary

Anaesthetics

There is considerable risk in these cases of post-operative chest complications, basal bronchitis and collapse This is minimized by intubation, avoidance of excess of ether, suction of any undue secretion from the air passages and by post-operative inhalations of carbon dioxide and oxygen at regular intervals for 48 hours

Transperitoneal approach

The most satisfactory approach is transperitoneal, through a paramedian incision extending from the costal margin to the pubis The post-parietal peritoneum overlaying the tumour is incised lateral to the colon (ascending or descending as the case may be) The colon is then reflected medially and complete exposure ensured The operator's first objective is to isolate and ligate the renal vein In dealing with the smaller and more mobile tumours this presents little difficulty Where the growth is more massive, some stripping and manipulation are a necessary preliminary, this should be done with the utmost gentleness and care. When isolated the vein is double-ligatured and divided, and the remainder of the pedicle also The enucleation of the tumour then proceeds, all surrounding tissues, perirenal fat, the adrenal, lymph glands and any adherent peritoneum or muscle being included The enucleation should be unhurried and thorough, and haemostasis meticulous The procedure is completed by replacement of the colon and repair of the peritoneum If thought advisable, a drain of corrugated rubber, introduced through a loin stab, may be left in place for 48 hours, but this is rarely necessary Finally the abdomen is closed in the usual way.

"Loin" approach

A transverse incision from the mid-line in front to the spine posteriorly gives a satisfactory enough access to most tumours, particularly those which are small and mobile It is, however, seldom possible to expose and ligate the pedicle without considerable preliminary manipulation of the tumour In fact the growth is usually delivered before the pedicle is clamped and tied For this reason, operation through the loin has been almost entirely superseded, though it has been employed in many of our cases in the past

Thoraco-abdominal approach

This affords perhaps the best access of all being particularly valuable in dealing with a high lying tumour suspected of fixation

Provision must be made to ensure the requisite positive lung pressure With the child in the lateral position the incision runs obliquely from the spine along the lower ribs and across the epigastrium to the mid line anteriorly The abdominal muscles including the rectus are cut across and the intercostal space usually that between the ninth and tenth ribs, incised throughout its length and the pleural and peritoneal cavities laid widely open the ribs being fully retracted and the exposure maintained by the use of a suitable self retaining instrument That devised by Denis Browne is excellent The diaphragm overlying the tumour having been incised in the line of its fibres the operation proceeds the same effort being directed to ligating the renal vein as early in the operation as may be feasible

Post-operative radiotherapy

This should be started as soon as possible Ideally the requisite plant should be in the same building as the theatre and the first dose may then be given immediately following completion of the operation Where this is not possible treatment should begin on the earliest day that the child s condition allows of the necessary journey

PARARENAL TUMOURS

The urological surgeon is often intimately concerned with the diagnosis and treatment of growths which arise in the adrenal gland or in the sympathetic chain on the posterior abdominal wall and it is therefore appropriate to include here a brief description of these tumours They are

adrenal cortical tumours
neuroblastoma and ganglioneuroma, and
phaeochromocytoma.

The last named is very rare and has been described in the section on hypertension (Chapter 4)

Adrenal cortical tumours

Tumours of the adrenal cortex form an interesting but comparatively rare group of cases They may present because of the local effects of the swelling or because of the endocrine disturbance consequent upon the excessive output of androgens Both features may well be evident in the same child but non hormonal tumours do occur and endocrine disturbance may result from a tumour too small to be clinically obvious In the latter group of cases it may be impossible to distinguish pre-operatively between tumour and hypertrophy of the gland

The hormonal disturbance is often described under the general term adreno-genital syndrome but the effects in male and in female are quite distinct In all cases of tumour the child has been normal at birth, but has later shown the characteristic virilizing changes by contrast, adrenal hypertrophy starting in foetal life may be responsible for female pseudohermaphroditism or in the male for precocity already evident at birth. In both sexes growth is abnormally rapid, muscular development excessive and centres of ossification appear considerably

earlier than in the normal child Fusion of the epiphyses is also premature so that despite the rapid growth in the earlier years, the child may be ultimately dwarfed compared with his fellows The blood pressure is usually raised In the male there is a precocious development of the external genitalia The penis is large and of adult appearance, pubic hair is plentiful and the skin tends to be thick and coarse The bodily form presents an impression which has been compared to "a burly brewers' drayman" The testicles do not partake in the development, however, and spermatogenesis is very rare, so that the term "precocious puberty" is not strictly applicable, nor do the boys adopt an adult attitude towards sex relations In girls the precocity is hetero-sexual the clitoris is enlarged, hair of male distribution appears on the body and perhaps on the face, and the feminine contours are lost

The excessive production of androgens is reflected in the increased excretion of 17-ketosteroids, but the degree of virilization is by no means proportional to the amount of androgen excreted, and it has been postulated that in some cases there must also be an excessive production of oestrogen Feminizing adrenal tumours are rare at any age and, like Cushing's syndrome, in which there is a more widespread disturbance of adrenal cortical function, are exceptionally rare in the early years of life

The tumour may be benign or malignant In the latter case it is often of rapid growth and presents as a solid mass with uneven surface in the upper loin and hypochondrium, fixed to its surroundings and displacing the kidney downwards In early cases, however, and with the benign tumour, the swelling is not clinically obvious and its localization may be in doubt Pyelography may demonstrate displacement of the associated kidney (*see* case V P , below) or the actual outline of the adrenal gland may be demonstrated in skiagrams following insufflation of oxygen into the perinephric space

Treatment

Where the tumour is malignant, the prognosis is naturally very poor A temporary regression of symptoms may be obtained by surgical removal of the growth, but recurrence and metastasis are the rule (*see* case A S , below) Happily this is not always so (*see* case V P below) In the less common benign tumours there is a reasonable prospect of permanent cure If only hypertrophy of the gland is discovered at operation, some regression of the virilizing changes may be obtained from a partial resection, but a recurrence is likely as a result of hypertrophy on the opposite side (*see* case J A , below)

Surgical excision of tumours should be attempted in all cases in which metastasis has not occurred, and very large and even fixed growths may sometimes, with due care and patience, be satisfactorily removed In simple hypertrophy removal of one entire adrenal and up to three-quarters or more of the other may have to be considered

CASE RECORDS

1 V P , female, aged 1 year 3 months Carcinoma of right adrenal—removal—cure

Referred by Dr Wyllie This little girl resembled in stature a child of 3 years, with a dusky red facies, excessive growth of pubic hair, huge nymphae and prominent breasts An adrenal tumour was suspected Under anaesthesia a small tumour was palpable

in the right adrenal area and the corresponding kidney was depressed as demonstrated pyelographically (*see* Fig. 108) At operation the right adrenal was removed with a small growth measuring 3 × 2 × 2 centimetres. The histology showed this to be a carcinoma. The child was given post-operative radiotherapy

Eighteen months later the child was well The pubic hair had vanished and her appearance was normal Now 8 years later she remains perfectly well

2. A S male aged 11 years. Carcinoma of adrenal—removal—temporary success only

Referred by Dr Wyllie. This child had been perfectly normal until 6 months before admission when he began to get very fat and suffer from spots on the face On examination he was a large fat boy with a round red face suggesting the brewers drayman type. The left kidney was palpable in the abdomen and appeared to be pushed down by a tumour above it The genitalia were well developed and there was a considerable



Fig 108 —Retrograde pyelogram showing downward displacement of right kidney by a cortical adrenal carcinoma Tumour removed from an infant girl aged 1 year 2 months. Alive and well (8 years)

growth of hair in the pubic region and in the axillae Blood pressure was 165/135 A skull skiagram showed a normal pituitary fossa. Intravenous pyelography confirmed the downward displacement of the left kidney and at operation a large adrenal tumour was removed This was reported as histologically carcinoma After operation there was a very striking and rapid improvement in his appearance and within a few months he was a normal boy Twelve months after operation the tumour and the clinical manifestations recurred and he died after 15 months.

3 J A. male, aged 5 years 9 months. Adrenal hypertrophy

Referred by Dr Wyllie The boy was normal at birth but at the age of 18 months his

mother noted that his genitalia were becoming abnormally large His growth then proceeded rapidly and he was considerably in advance of his elder brother No tumour could be felt on clinical examination and the intravenous pyelography was normal 17-ketosteroids=18 4 milligrams excreted in 24 hours A laparotomy was performed the left adrenal felt normal but there was a lump at the upper pole of the right kidney This was approached at a later operation from the loin The supra-renal was found to be enlarged but there was no evidence of tumour A portion of the gland was resected There was perhaps a slight improvement after the operation, but 4 years later the boy is considerably advanced in appearance, his bone age is estimated at 16 years (chronologically aged 9), and ketosteroid excretion is 22 milligrams per 24 hours The penis is large but the boy's mentality is normal for his age It is felt that in this case more extensive adrenalectomy should have been done

Neuroblastoma and ganglioneuroma

These two tumours both arise from primitive nerve cells, in the former, differentiation is poor and the growth extremely malignant in the latter the cells are more mature and the course correspondingly benign No hard and fast distinction is possible, however, and intermediate forms are not uncommon, while maturation of cells in a tumour initially malignant has been observed

Neuroblastoma and ganglioneuroma are not found in the central nervous system, but in connexion with the autonomic ganglia and with the medulla of the adrenal glands Willis (1948) estimates that one-third of all these tumours arise from the adrenal and another third from the abdominal sympathetic system, in both of which situations they are of importance to the urologist The primitive nerve cells from which they are formed (sympathogonia) migrate from the central nervous system into the retroperitoneal area where differentiation into sympathetic ganglion cells and adrenal medullary cells takes place Migration is occurring during a long period of foetal and perhaps of post-natal life, the tumours are formed only from these undifferentiated cells and are therefore truly embryonic in character (Willis, 1948) They are therefore found predominantly in young children, though the more benign varieties may not present until later in life

Pathology

Histologically the most malignant of the neuroblastomas consist of small round or polygonal undifferentiated cells arranged without any distinctive pattern, and there can be little doubt that many of the tumours formerly diagnosed as retroperitoneal sarcoma or lymphosarcoma come within this group In some of the tumours, the cells are arranged in characteristic rosettes with central differentiation of young nerve fibrils When seen, this appearance suggests an early stage of differentiation Areas of necrosis and haemorrhage are common and calcification may occur The tumour is commonly of rapid growth and metastasizes early Infiltration of the retroperitoneal plane, with involvement of the adjacent viscera, has occurred in most cases by the time they present for treatment The distribution of metastases is variable, there may be massive involvement of the liver (Pepper's syndrome), but more often the secondary deposits are seen farther afield particularly in the skeleton, notably in the skull and long bones Sir Robert Hutchison (1907) described the group in which cranial and orbital deposits, associated with exophthalmos, were the prominent features (Hutchison's syndrome) It was

formerly thought that tumours of the right adrenal were always associated with the Pepper type and those of the left with the Hutchison type of dissemination The truth is not so simple Metastatic deposits in the liver (Pepper) are certainly much more commonly associated with a primary growth in the right adrenal but with the Hutchison picture the primary growth may be found on the right or left indiscriminately The bone metastases may be purely osteolytic but sclerosis is not uncommon and the appearance of the spine and pelvis sometimes resembles Paget s disease Wyatt and Farber (1941) comment upon the frequent symmetrical arrangement of bone secondaries Willis (1948) has demonstrated that the onion skin lamination of tumours of long bone, previously regarded as characteristic of Ewing s sarcoma, may also result from neuroblastoma secondaries

In the ganglioneuromas cellular differentiation has proceeded further than in the neuroblastomas nerve cells may be easily distinguishable multi nucleated cells resembling the normal ganglion cells are not uncommon and a tangle of nerve fibres may be found A case in which the histology was originally neuroblastoma, but which underwent spontaneous evolution to the ganglioneuroma appearance, was first reported by Cushing and Wolbach (1927) and many subsequent authors have been impressed by the possibility of this type of maturation (*see* Fig. 111) By contrast, a tumour which has been apparently quiescent for some years may suddenly take on a malignant character Ganglioneuroma may be an entirely benign and encapsulated tumour of very slow growth but local infiltration and fixation to adjacent organs is common in the intermediate forms

Clinical features

Neuroblastoma occurs with approximately the same frequency as nephroblastoma and in much the same age group that is under 5 years The sexes are equally affected Ganglioneuroma is very much less common and more likely to be seen in older children In tumours of the adrenal area the malignant variety unquestionably predominates The first sign to be noticed is often the abdominal swelling, but general symptoms of ill health lethargy loss of appetite pallor and wasting, are prominent features and tend to appear earlier than in the renal growths There is often a complaint of pain and it would seem that these growths involve a retroperitoneal somatic nerve at an early stage In some cases secondary deposits notably those of the Hutchison type may be the earliest evidence of the condition Stern and Newns (1937) have pointed out the significance of bone pains as an early indication of metastasis

In tumours arising from the adrenal area the abdominal swelling may be indistinguishable from a nephroblastoma, particularly of the extrarenal type The neuroblastoma, however is more deeply placed, is usually more central and is apt to cross the mid line The tumour feels more nodular fixed and of indeterminate limit. Distinctive outlying nodules are not infrequently present In tumours arising from the sympathetic in the lower lumbar region the distinction from renal growths does not present the same difficulties the neuroblastoma is close to the mid line and fixed to the posterior abdominal wall Calcification may be seen on a plain skiagram more commonly in the neuroblastoma than in the renal embryoma It may take the form of a uniform stippling throughout the tumour area or there may be irregular confluent masses The pyelogram often affords more reliable



Fig 109

FIG 109—Excretion pyelogram showing displacement without distortion of left pelvis in a neuroblastoma of the adrenal Note the calcification Girl aged 5 years

(By courtesy of Edward Arnold & Co)

FIG 110—Retrograde pyelogram from an infant girl aged 10 months showing displacement of the left ureter by a pelvic neuroblastoma

During an attack of enteritis a lump was discovered *in the left pelvis rounded firm* and fixed to the posterior wall At operation a solid tumour the size of a tennis ball and purple in colour was enucleated from the root of the mesentery of the lower pelvic colon The histology was that of a neuroblastoma of high malignancy The infant's recovery was uneventful and so far she remains well



Fig 110

evidence In the adrenal tumours the renal pelvis is characteristically displaced and rotated downwards and outwards without suffering any significant distortion (*see* Fig 109) By contrast in renal growths even where displacement is the most prominent feature some definite distortion of the calyceal pattern is the rule Neuroblastomas arising below the kidney commonly displace the ureter (*see* Fig. 110) and may compress it causing a hydro-nephrosis Radiological investigation of the bones and lungs should always be undertaken

Treatment

Surgical excision must always be the primary aim of treatment, but local fixation or metastasis all too frequently renders this impossible The operative procedure is as for renal embryoma and the thoraco-abdominal route should be employed The



Fig 111 —Excretion pyelogram from a girl of two years showing marked calcification and dilatation of the renal pelvis

This child was operated on for a neuroblastoma which was stated to be very large and which proved inoperable. There has been remarkable post-operative shrinkage and the child is in vigorous health 4 months later No biopsy was made and the diagnosis must therefore be in doubt. Nevertheless we believe this to be an instance of regression of a neuroblastoma.

tumours are often excessively friable, and where the extent and attachments of the growth appear very formidable it is better to abandon the operation rather than risk the extrusion of tumour substance and haemorrhage, which will result from rupturing the capsule during heroic attempts at removal A biopsy however should then be taken The more benign varieties and the ganglioneuromas may be encapsulated and removed with comparative ease Neuroblastomas are radiosensitive though they never shrink with the same rapidity as do most of the Wilms tumours, and the ultimate results are disappointing. At the Hospital for Sick Children all those tumours which have arisen within the adrenal itself have been

fatal, but the possibilities of maturation and of spontaneous shrinking have been emphasized by Farber (1940). Tumours arising in the extra-adrenal elements of the sympathetic chain, however, would seem to be somewhat more hopeful since, among this comparatively small group, there are a few survivors. Many chemotherapeutic agents reputed to control malignant disease have been tried out on the neuroblastomas, but have so far been, in our experience, uniformly disappointing.

CHAPTER 14

CONGENITAL EXTERNAL ANOMALIES

HYPOSPADIAS

THE embryology concerned in this deformity has been fully outlined in Chapter 6. The anomaly represents an arrest of development resulting in failure of closure of the urethral folds of varying degree Thus the terminal orifice may be. (1) in the coronary sulcus (glandal or coronal type) (2) at any point along the shaft of the penis (penile type) and (3) in the perineum in which case the labioscrotal swellings have also failed to unite (perineo-scrotal type) In these extreme cases the existence of a vaginal pouch may render determination of sex difficult (*see* Chapter 16)

Associated anomalies in canalization of the urethral plate determine various appearances which will be seen on the glans There may be a complete gutter or a blind pit at the site of the normal meatus. Not infrequently blind lacunae which as can be very simply demonstrated by drawing the two wings of the hood together may be a centimetre or more in depth, occur along the course of the glandal urethra.

The characteristically hooded appearance of the prepuce is due to the ventral defect caused by failure of the lateral folds to close and their resulting retracture, on the under-surfaces of the glans when the prepuce assumes a normal aspect except for the ventral incision

Most important from the point of view of treatment is the ventral curvature of the penis (chordee) which in some degree almost always accompanies the deformity

In the simple coronal type the glans is often angled ventrally a deceptive appearance due to absence of substance on the under-surface and not to any remediable tissue contracture In the penile and perineo-scrotal types chordee, so commonly present, is attributable to the short urethra, failure of the corpus spongiosum which is represented by a fibrous cord, and also to fibrosis along the ventral surface of the corpora cavernosa The tethered penis cannot grow until it is adequately freed The first step in reconstruction therefore consists in separating the urethra from its forward attachment, displacing it backwards so that the true length of the defect becomes manifest, and at the same time carefully disposing of all associated anchoring fibrous tissue In this way the penis is straightened and expansion and growth made possible.

The terminal urethral meatus particularly in the coronal and penile types, is very often minute and in infancy may be difficult to find until demonstrated by the passing of urine It may be necessary to dilate the opening with a probe or even to carry out a formal meatotomy (*see* Fig. 112)

Since the developmental error is in all cases distal to the urinary sphincters, there is no inherent defect of bladder control

Treatment

Infancy

The problem usually presents first in infancy, when the points to be determined are (a) the degree of the deformity, (b) the adequacy of the urethral orifice, and possibly, in the perineo-scrotal type, (c) the sex of the child

A "pin-hole" meatus will require immediate operative relief, either by dilatation or meatotomy, since some degree of retention may be caused

In the perineo-scrotal cases the possibility of female pseudohermaphroditism should always be borne in mind The existence of a vaginal orifice and absence of testicles are always suggestive The infant should be kept under observation



FIG 112 —Penile hypospadias in a child aged 4½ years

until further investigations become feasible (for instance, utero-salpingograms, 17-ketosteroid estimations) and for obvious reasons these should not be long delayed (*see* Chapter 16)

Decision as to operative treatment

Coronal (*type* 1).—In this minor degree the penis is straight and properly developed Normal function is assured, the urinary stream will be directed forwards in a natural manner, and there will be no wetting of trousers In later life the defect is no deterrent to the founding of a family

The ventral deficiency of the glans (pseudochordee) which is a more obvious disfigurement in some cases detracts in no way from function There seems there fore no adequate reason for any serious operative interference in cases of this type moreover damage to important local sensitivity is likely to make such attempts positively harmful

The cosmetic appearance may be greatly improved by an appropriately modified circumcision in later childhood and any blind pits in the glans for example may be laid open at the same time if thought necessary we would however deprecate any procedure designed to restore the glandal segment of the urethra

Penile and perineo-scrotal (*types* 2 *and* 3) —In many of the cases in this group the urethra opens well forward on the penis though short of the coronal sulcus and there may often be some degree of true chordee the urethra being in reality shorter than it appears It is useful to call this variety pre-penile to distinguish it from the coronal type with which it is sometimes confused In the true pre penile case the urinary stream is likely to be directed downwards and growth of the penis hampered Reconstructive surgery is therefore usually desirable

In all the more severe degrees of penile and penoscrotal deformities operative treatment is required In most, initial correction of the chordee is necessary followed later by reconstruction of the urethra

Age of operation

It is generally recognized that restoration of normal masculine micturition must, if possible, be achieved before the school age, say 5 to 8 years For psychological reasons it is obviously important that the boy at school should pass urine like his fellows

The preparatory first stage, the correction of the chordee is undertaken as early as possible preferably at 18 months to 2 years so that the fullest opportunity may be afforded for expansion and growth of the penis The final reconstruction may then be planned for the 5th to 7th year

Operation stage I

Assuming that chordee is present the first necessity is to straighten the penis Though this is not unduly difficult care is necessary if the best chance is to be given to the subsequent plastic procedure

A transverse incision is made on the ventral aspect, well forward of the urethral orifice and close to the glans This incision should be long enough to expose both corpora cavernosa laterally and extended into the prepuce on either side (*see* Fig 113) By wide undermining with fine blunt pointed scissors the under surface of the penis is laid bare and the contracted fibrous remnant of the corpus spongiosum is exposed This is freed and dissected from before backwards and the urethra then gradually recedes towards the perineum demonstrating the full extent of its shortening. When the urethral orifice sits comfortably without tension the fibrous tag is trimmed off All further lateral fibrous bands are now divided, great care being taken not to injure the underlying corpora cavernosa The original transverse incision is now approximately longitudinal and it is further lengthened by splitting and undermining the incised prepuce freely The incision is finally closed vertically by sutures of fine unhardened cat gut passed on atraumatic

needles. Any possible tension is relieved by making a mid-dorsal incision through the skin of penis and prepuce, spreading the edges widely as advocated by Browne (1950). The penis may be wrapped in ribbon gauze, soaked in acriflavine and paraffin, or strapped to the pubes with Elastoplast over a suitable pad to ensure dorsiflexion.

Following the successful completion of stage I, normal growth of the penis is to be anticipated, and as much time should be allowed for this as possible In certain cases hormones may help (testosterone pellets, 200 milligrams, one or two, according to age, buried in the subcutaneous fat of the abdominal wall or buttock)

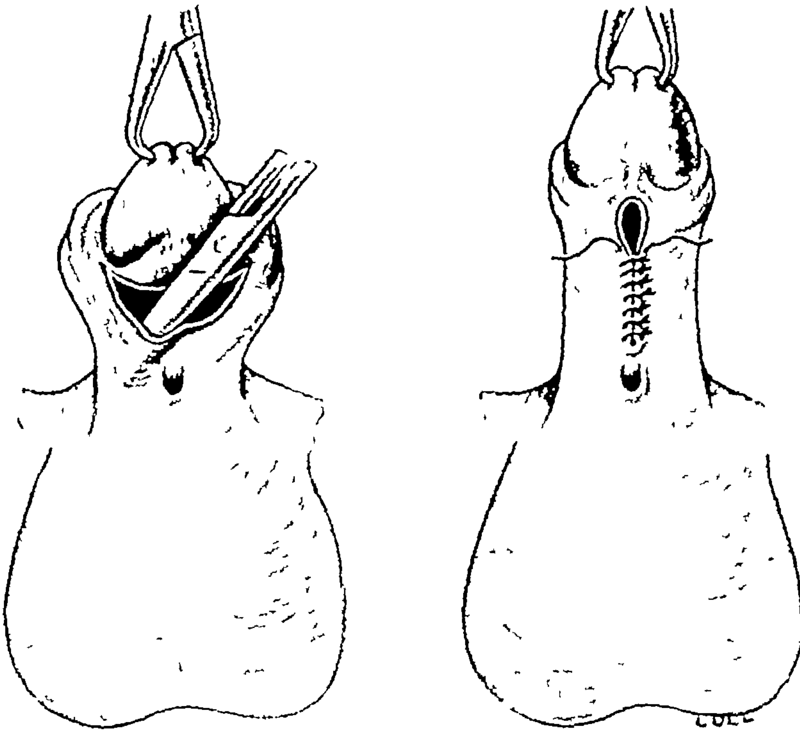


FIG 113 —Hypospadias Operation Stage I Repair of chordee

Operation, stage II, Age 5–7 years

Many ingenious operations have been devised and advocated in the past for refashioning the urethral tube Some which have been described were quite unsuitable for small children, and by and large the results left much to be desired The pioneer work of Edmunds, and more recently of Denis Browne, has completely changed the prospect for the boy unfortunate enough to be afflicted with this deformity Those of us who have been privileged to see the modern Denis Browne operation in action, and to practise it under his guidance, cannot but feel that this operation will come to supersede all others, at any rate in childhood. The technique is not too difficult for the "average competent surgeon" to master, and the immediate results are remarkably satisfactory. Any breakdown of the suture line, or formation of fistulae, is rare (3 instances in 50 operations (Browne,

1949)) The new urethra is of adequate size and appears to remain so It may of course be said with truth that no final judgement can be given of any such operation until the patient has reached adult life, and the evidence as yet only extends for 4 or 5 years All that can be said is that so far there is no sign of post-operative contraction whenever Browne s technique has been faithfully followed There are of course possibilities of error in the performance of the operation and no written description however adequate, can take the place of visual demonstration We would urge any surgeon before embarking upon the operation to see it done, either by the master or a competent disciple The following account of the operation is based entirely upon Browne s own description as published in *Techniques in British Surgery*. Opportunity is taken to stress those points in the procedure at which mistakes are apt to be made by the inexperienced. Browne s own description is extensively quoted

Before embarking on the operation at all it is essential that the ground should have been satisfactorily prepared that is that the result of stage I (relief of the chordee) should be completely successful, the penis straight and reasonably well grown the skin of the under surface smooth and supple, and the scar of the original incision not unduly noticeable. Such a desirable result does not always follow the initial operation and there should be no hesitation whenever it seems necessary in carrying out a further preliminary trimming by excising scar tissue

In those uncommon instances where no preparatory operation for correction of chordee is necessary the foregoing does not, of course, apply

It is important also to ensure that the ectopic urethral meatus is of adequate size, since this must not constitute a narrow point when it is incorporated in the new urethra. Browne very wisely stresses the need that the over all calibre of the new urethra should be adequate to permit of the introduction of a cystoscope if it should ever be necessary It is important therefore before embarking on the final stage to assure one s self on this point. If the urethral outlet seems too small it should be fully enlarged by a meatotomy if necessary and further operation postponed for the moment

Principle of the Denis Browne operation

Before describing the further steps of the operation it is pertinent to enunciate the principle upon which it is based Denis Browne writes as follows, speaking of post-operative urethral fistulae in general They differ from similar fistulae elsewhere in the body in being lined with epithelium indistinguishable from that of the surrounding normal surface and in not being encircled by scar tissue with its inevitable tendency to contract. In consequence they have no tendency to close spontaneously It will be seen that these fistulae have just the properties desired for an artificial urethra and it is on this curious immunity of the penile skin and subcutaneous tissue from keloid or contraction that the recommended operation is based It consists, in fact in the deliberate formation of a fistula to end all fistulae A simple strip of skin when buried beneath the surface is enough to form this permanent fistula a hollow tube is quickly formed by the growth and rolling up of this strip and the urethral lumen can in this way be prolonged up to the tip of the penis by a comparatively simple operation

The operation

Urethral drainage of the bladder—Diversion of the urinary stream from the operation area is essential, and Browne has emphasized the defects of supra-pubic drainage Even with continuous suction, escape of urine into the urethra occurs sooner or later, causing the child great pain, flooding the wound, and leading almost inevitably to breakdown of suture lines Urethrostomy drainage of the bladder avoids this drawback and has proved eminently satisfactory in practice Browne's method of introducing the drain is as follows: A Malecot catheter is selected of such a size as will be a reasonable fit Mounted on a sound and well lubricated, this is passed into the bladder. When urine flows, the sound is withdrawn slightly and turned through 90 degrees, so that the tip stands out in the perineum The sound is then held by an assistant and the operator fixes the tip with his left finger and thumb, exactly in the mid-line of the perineum With the diathermy knife, a small incision is made and deepened until the rubber of the catheter is seen The point of the sound is then insinuated into the wound so that the catheter can be readily grasped by forceps The hold must be adequate and firm lest the catheter slip back on withdrawal of the sound, which is the next step The catheter is now carefully manoeuvred backwards along the distal urethra, until it emerges through the perineal incision If bladder urine is then found to be still flowing freely, the catheter is spigoted and "stitched to the skin by two linen sutures, tied tightly round the tube and loosely in the skin so that they do not cut out. If this is not done, even the best behaved child may pull the catheter out, particularly when half asleep "

The whole manoeuvre sounds and looks easy, but there are pitfalls In incising the perineum it is absolutely essential to keep strictly to the mid-line, otherwise unpleasant haemorrhage from the corpus veins is met with In one of our own cases this proved a serious mishap causing us considerable anxiety for some days

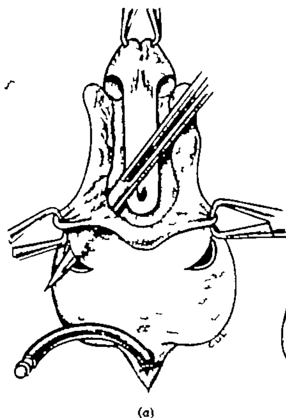
It is important to keep the urethral incision as small as possible, that is, the catheter should completely fill it This ensures rapid closure of the fistula after subsequent withdrawal of the catheter

During the manipulations, the catheter may easily be withdrawn from the bladder If urine fails to flow, the catheter must be manipulated into the bladder beyond all doubt before it is fixed by suture

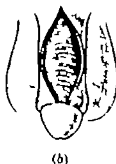
"The end of the tube is later connected up to a reversed Wolff's bottle, giving a suction of about a foot of water, which will be enough to keep the bladder empty without discomfort "

Fashioning the new urethra—An incision is made as outlined in Fig 114 (*a*), encircling the urethral orifice and extending above to the glans The strip of skin enclosed should be of adequate width One of the common mistakes of inexperience is to make the strip too narrow, and this will result in an unsuccessful narrow urethra On the other hand, it must not be needlessly wide or the covering flaps are likely to prove inadequate

The lateral flaps are now widely and freely undermined by gentle dissection using fine scissors on the Mayo principle Special attention must be paid to this undermining below the urethral opening in the direction of the perineum So far as possible the lowest point of suture, when the flaps are united, should lie well forward of the old urethral opening When this part of the undermining involves



(a)



(b)

FIG 114.—Hypospadias. Denis Browne operation
(a) Incision and fashioning of flap—perineal urethrostomy drainage tube in position site of stab drain in scrotum indicated,
(b) dorsal incision for relief of tension.

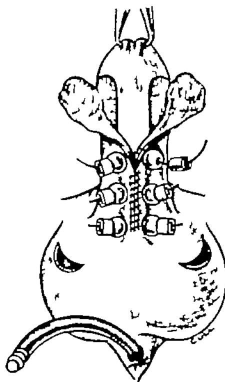


FIG 115.—Hypospadias. Denis Browne operation Suturing of flaps

the scrotum, stab wounds are made on either side at the deepest point of the post-urethral pouches so formed to allow ready escape of blood which might otherwise collect in the scrotal tissues and possibly lead to infection (*see* Fig. 114 (*a*))

The whole undermining process must leave the flaps so free that they "come easily together like the sides of a loose double-breasted coat"

Next "a triangular piece of glans is made raw on either side of the situation of the new meatus, this being most easily done by shaving the skin off with sharp scissors"

Dorsal incision—"The necessary relaxation for the joining of the lateral flaps over the urethral strip is gained by a simple longitudinal cut all the way down the dorsum of the penis (Fig 114 (*b*)) This may be made before or after the stitching *of the flaps and is left to epithelialize itself It does this with the characteristic* local lack of contraction."

Tension sutures—The flaps are now sutured The following is Browne's description "These sutures should be what I call the 'double-stop' type Their hold is due first to a stop on the skin, and second to a stop on the suture itself When employed on the penis the skin stop is formed of a glass bead about $\frac{1}{4}$-inch in diameter, though elsewhere buttons or plates of appropriate size may be used. The suture stop is formed of a small section of soft aluminium tubing, which is crushed where needed by strong forceps, an old artery forceps with the jaws shortened to 1 inch or so will do admirably I have found aluminium the only suitable metal for this purpose, lead is too soft and slippery, and most other metals too hard (Fig 115)

"These sutures are inserted as shown in the drawing, taking in a generous bite of the tissues on either side, *but great care must be taken not to draw them too tight** as that would produce necrosis of the delicate skin in their grasp The beads should be brought together, and then the one to be fastened withdrawn at least $\frac{1}{8}$-inch before the suture stop is clamped down"

Skin sutures—"The skin edges of the cleft are now brought together with extremely fine and carefully applied sutures of cat-gut, taking the minutest possible grasp of their edges There should be no tension whatever upon this line, the sutures merely having the task of adjusting and preventing inversion". (*See* Fig 115)

"Finally the newly constructed ventral surface of the penis is pulled well up on to the glans, so that its raw undersurface is against the raw patches made on either side of the new meatus, and there it is sutured firmly into position"

Dressing—"This is simply a spray of penicillin and sulphonamide powder, the operation area being otherwise left open to the air This powder, mixed with exudate, produces an antiseptic scab of the kind approved by Lister, and healing proceeds very well under it"

After-treatment—"Penicillin injections are kept up for the first week after operation, the aseptic healing usually obtained by them being well worth the extra discomfort The tension sutures are removed at the end of the week by cutting between the two elements of the stop As the beads by this time are usually

*The italics are ours

embedded more or less deeply in swollen and sensitive skin the advantages of this method of cutting are obvious. The drainage of the bladder is kept up for ten days and the opening in the perineum usually closes within three to five days later

ECTOPIA VESICAE AND EPISPADIAS

Anatomy

Ectopia vesicae and epispadias are most important types of a group of deformities which have in common some deficiency in the mid line of the infra umbilical abdominal wall In the typical case of ectopia vesicae the defect is complete, the anterior bladder wall absent and the vesical mucosa prolapses through the abdominal wall defect The umbilicus which is rather lower in the abdomen than normal is represented by a scarred-area at the apex of the bladder occasionally with an associated ventral hernia At the base the trigone can be seen and in the male in whom this deformity is somewhat more common the posterior urethra is laid open between the widely separated pubic bones so that the verumontanum can usually be identified The penis is short thick-set and uptilted, the crura being widely apart on the ischio-pubic rami The urethra forms a groove on the dorsal surface of the penis lying above the corpora cavernosa. The prepuce is split dorsally but has a normal frenum ventrally and the faint median raphe on the under-surface of the penis can be made out. The scrotum is normally fused but may be empty (*see* Fig. 116)

With lesser degrees of the defect the umbilicus lies above the bladder which though deficient anteriorly forms a definite sac within the abdomen The bladder itself may be covered in leaving a defect only of its sphincters and of the urethra —the common type of epispadias (*see* Fig. 117) In still milder forms the sphincters are present and preserve continence, leaving only the characteristic deformity of the penis Occasionally as has been pointed out by Thompson (1937) there may be a few strands closing over the bladder neck even with the fully ectopic bladder

In the female with complete ectopia a vaginal orifice can usually be found below the bladder at about the level of the pubic rami the vagina is short and the cervix near the surface. The clitoris is divided and formed below and posterior to the urethra (Fig 118) Epispadias in the female is the only form of this anomaly which is liable to be overlooked though it cannot be mistaken by anyone familiar with the condition The urethral meatus more or less normally placed, appears as a wide transverse slit and the urethra itself is so short and wide that the trigone may be visualized by a simple retraction of its margins. The clitoris is bifid and each half lies lateral or postero-lateral to the urethra while anteriorly a broad area of glabrous skin stretches upwards to a deficient mons veneris (*see* Fig. 119) The symphysis pubis is commonly un united but the degree of bony fusion bears no relation to the integrity of the urinary sphincters and a normal symphysis may be associated with complete incontinence

The separation of the pubic bones is associated with some instability of the pelvic ring which often produces a curious waddling gait. The weakened condition of the musculature of the pelvic floor may be evidenced by recurrent rectal prolapse



Fig 116

FIG 116 —Ectopia vesicae Male aged 4 years

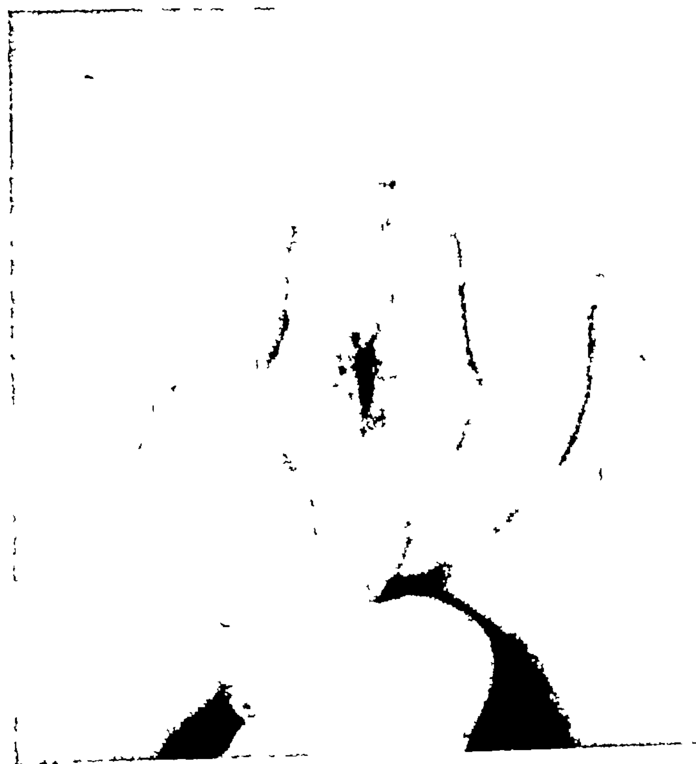


Fig 117

FIG 117 —Epispadias Male aged 6 years (continent)

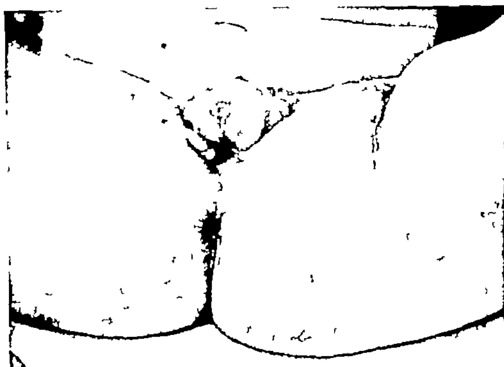


FIG 118.—Ectopia vesicae. Female aged 1 year 10 months (note position of umbilicus)

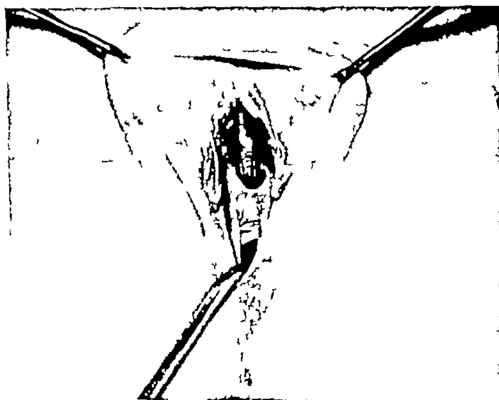


FIG 119.—Epispadias. Female aged 5 years.

Among the minor forms of the ventral mid-line defect are the "pubic umbilicus" (an umbilicus placed low in the abdomen but unassociated with bladder disorder), and perhaps the short dorsal para-urethral ducts which lie immediately beneath the skin of the dorsum of the penis and have an epispadias-like opening on the glans

Embryology

The embryology of the mid-ventral defect has not yet been satisfactorily worked out, but the final solution of the problem will probably be upon the lines suggested by Wyburn (1937) This author believes that in the very early embryo (embryonic disc stage) there is a primitive contact of endoderm with ectoderm in the region between the allantoic diverticulum of the yolk sac and the tail bud (which represents the caudal extremity of the primitive streak), elsewhere, even in the earliest embryos, some mesoderm intervenes between the two primary germ layers With the formation of the tail fold and of the hind gut, this area of contact between ectoderm and endoderm comes to lie between the tail and the body stalk, and the caudal part of it forms the cloacal membrane Normally the cranial part of the area is invaded by mesoderm from either side, and the fusion and growth of this mesoderm is responsible for the separation of the umbilical stalk from the region of the cloaca and for the formation of the infra-umbilical abdominal wall, the genital tubercle, the symphysis pubis and the anterior musculature of the bladder Should failure of this mesodermal invasion occur in whole or in part, the cloacal membrane consisting of the single apposed layers of ectoderm and endoderm will reach from the tail to the umbilicus, and when it ruptures the bladder will be left widely open Such an origin for ectopia vesicae entails a deviation at an extremely early stage at which even the normal processes of development are far from clear The occasional association of ectopia with gross deformities of the gut and of the hind end of the body also suggests a very early deviation

It must be recognized, however, that even in complete ectopia the infra-umbilical mesoderm, though misplaced, is not entirely deficient and the genital tubercle and its derivatives are present The under-surface of the epispadiac penis shows evidence of a median raphe under which may be felt a fibrous cord, suggesting that some process similar to the normal fusion of urethral folds has occurred Moreover, although the bladder and posterior urethra have the appearance of the normal passages laid open, the penile urethra lies dorsal instead of ventral to the corpora cavernosa, a situation which demands a perversion of the normal development rather than a simple failure of mesodermal fusion

Ectopia vesicae

Clinical picture

Ectopia vesicae is one of the most grievous of congenital deformities and reduces an otherwise normal child to a pitiable state of misery and loneliness The exposed vesical mucosa is acutely tender to the touch and the unmanageable incontinence leads to a severe excoriation of the surrounding skin The waddling gait produced by the instability of the pelvis is aggravated by the desire to avoid friction to the raw area, and recurrent prolapse of the rectum may place one more handicap upon this already overburdened child

With the passage of time, particularly if the urine has been diverted the mucosa of the upper part of the bladder tends to undergo squamous metaplasia and to take on the appearance of skin Where it is sodden and infected, however the mucosa looks angry and oedematous and microscopy may show glandular metaplasia If the condition is allowed to persist to adult life there is a certain danger of neoplasia complicating the chronic infection

Treatment

The object of treatment is to attain continence to eliminate the tender exposed mucosa and to produce an approximation to the normal appearance of the genitalia Continence can almost always be achieved following a period of training by transplantation of the ureters into the bowel and our experience leads us to believe that this operation is not attended by serious complications in childhood and that after it renal function remains surprisingly good Subsequent plastic procedures are, of course undertaken but it cannot be pretended that a child without a bladder is as good as a child possessing one, and the small boy who has of necessity to adopt the sitting position for micturition is not likely to escape the awareness of his inferiority or the ridicule of his fellows Reconstitution of the bladder is therefore the ideal of surgical ambition and where there is a prospect of a capacious organ attempts at reconstruction may be justifiable It cannot be claimed, unfortunately that a thoroughly satisfactory method of achieving continence has yet been devised In some cases the bladder area is so small that no hope can be entertained of forming an organ of adequate capacity in many however the construction of a saccular bladder presents no particular difficulty though it is wiser to close the defect in the abdominal wall by the rotation of lateral flaps rather than by simple mid line suture A tubular urethra can be formed by burying the mucosal strip and approximating the superficial tissues, but there is apt to be some trouble in closing the fistula at the junction of bladder and urethra It is, however in the final stage—the achievement of continence—that most reconstructions have failed, and reports of successful cases are extremely rare Young (1942) was able to form an adequate sphincter in a girl by excising the anterior part of the reconstructed bladder neck and by wrapping the surrounding tissue about it Michon (1948) reports a case in a boy in which a gracilis transplant encircling the newly formed urethra maintained a continence despite complete fibrosis of the muscle, and another a girl in whom the bladder was completely closed off the urethra being subsequently formed by the tunnellization method described by Marion for cases of vesico-vaginal fistula Since the latter method requires repeated dilatation throughout life it is unlikely to achieve great popularity

Until a more satisfactory reconstruction has been devised therefore the treatment of choice in cases of ectopia vesicae is transplantation of the ureters This operation will normally be postponed until after the period of physiological diuresis that is until 18 months to 2 years of age but there is nothing to be gained by delaying it beyond this time Until the operation is performed everything must be done to mitigate the effects of the ammoniacal decomposition of the urine upon the surrounding skin Constant wetting is unavoidable so that frequent changing of napkins and cleansing of the area are obligatory Whenever

possible the skin should be freely exposed to the air, it may be protected by smearing with vaseline or with a buffered gel (P.V A Jelly) * In certain cases the preparation Siccolam (B D H Ltd) is a useful application

Control of the *B ammoniagenes* factory in the bowel should be maintained as far as possible by breast feeding and bowel antiseptics Where rectal prolapse is present it should be treated before the transplant is undertaken, and for this purpose a subcutaneous encircling cat-gut stitch at the anus is often useful Occasionally very large inguinal herniae have to be repaired during the early months of life

The operative technique of ureteral transplantation described below has caused no anxiety and has been free from any immediate mortality during the past 10 years

Technique

Preliminary sterilization of the bowel is achieved by sulphathalidine or streptomycin given for 3–4 days The right ureter is transplanted first, the left afterwards within 2–3 weeks A paramedian incision is made immediately above the vesical mucosa, the peritoneum opened and the ureter isolated, a process which may be facilitated by the preliminary passage of a ureteric catheter The ureter is divided as low down as possible, the lower end tied off and the peritoneal defect partially sutured The proximal end is then brought to the sigmoid as low down as is convenient, a submucous channel is made by incising the muscularis of the bowel, the edges of which are then closed over the ureter by stitches which pick up the muscle of the ureteric wall The free end of the ureter is then drawn in to the colon through a stab wound at the end of the submucous channel, and finally the whole junction is embedded by Lembert's sutures and covered by completing closure of the original peritoneal incision The abdomen is closed without drainage The urine rarely appears in the bowel before the third or fourth day after this operation, but no harm results from this delay The second ureter is transplanted two weeks later Some days, perhaps some weeks, of training are required to achieve full continence after the transplantation

The exposed vesical mucosa is dealt with some time later Diathermic coagulation may be adequate and following it the area occasionally becomes satisfactorily "skinned over", but should this fail, or should there be a large hernia-like defect, the bladder mucosa should be excised, and the raw area covered by lateral flaps rotated medially Finally the epispadiac deformity of the penis should receive attention a simple adaptation of the Denis Browne procedure for hypospadias will close the urethral groove but it is very difficult to correct the uptilting of the penis, which presumably results from the unusual origin of the crura The bony

*The preparation of P V.A Jelly is as follows

Buffer solution		
Potassium hydrogen phthalate		10 2 g.
N/1 sodium hydroxide		23 9 ml
Chlorocresol		1 0 g
Water to		1000 0 ml
Solution A	Congo red	1 g
	Buffer solution to	100 ml
Solution B	Polyvinyl alcohol	10 g
	Buffer solution to	100 ml

Equal volumes of solutions A and B are mixed together at 50°–60 C. (*p*H 5 0 5 2)

defect seldom requires specific treatment but the gait may be improved by training so far orthopaedic procedures designed to secure union of the symphysis have not been successful.

The long term results of ureteric transplantation have on the whole been very satisfactory though a slight degree of upper tract dilatation is the rule and it has occasionally been more severe A mild hyperchloraemic acidosis may rarely be encountered following transplantation and regular dosage with alkalis is then advisable

Epispadias

Although incontinence is the rule in cases of epispadias it is not quite such an unmanageable incontinence as with ectopia vesicae and the acutely tender exposed mucosa is absent. While the child is lying down the bladder may hold quite an appreciable volume of urine which floods out, however on standing up

The chances of attaining continence in cases of epispadias are rather greater than after reconstruction of the bladder in cases of ectopia, and repeated attempts should always be made to build up the urinary sphincter before resorting to ureteric transplantation The mucosal strip should be freed from the skin and from the region of the pubic bones so that it can drop back to a more normal position The superficial tissues may be approximated in front of the mucosa, which is left to form itself into a tube as in the Denis Browne operation for hypospadias A wedge of mucosa may be excised from the anterior part of the bladder neck, and the adventitial layer on either side drawn tightly together in this region The operation is more easily performed where the symphysis is un united and the whole length of the urethra can be displayed, but there should be no hesitation in cutting the symphysis where union has occurred In the female cases there is the additional problem of a very short urethra and in some there can be little hope of restoring bladder function

ANOMALIES OF THE URACHUS

The growth of the urogenital sinus in the early embryo carries the apex of the bladder up to the umbilical stalk, and the allantois is lost. The urachus is formed by a narrowing down of the upper extremity of the bladder not from the allantois, and it is lined by epithelium similar to that found in the bladder In the normal course of events the urachal canal is closed at both ends and is itself almost obliterated Cyst formation or failure of closure may however occur and the following anomalies result

Patent urachus (Fig. 120 (A)) —The bladder communicates with the umbilicus through a narrow channel and urine discharges from the umbilicus in varying degrees—it may be continuously or only at intervals Recognition is not difficult unless the discharge is very slight The orifice is usually a skin lined track, though in some the mucosa pouts and may be mistaken for granulation or a polyp The injection of Lipiodol into the tract or a cystogram will confirm the diagnosis in doubtful cases

Fig. 120 (B) represents a rarer type of this anomaly

Urachal cyst (Fig. 120 (C)) —The track has closed from the bladder below and from the umbilicus above though the latter may be incomplete a minute fistulous

track allowing a certain leakage and so explaining delayed appearance of the cyst The cyst forms a rounded lump in the mid-line below the umbilicus It is attached to the deep layer of the abdominal wall in front of the peritoneum Ladd and Gross (1941) emphasize the risk of infection, and mention one of their cases in which a calculus formed in the urachal tract in a 7-year-old boy A cystogram may show a compression deformity of the fundus of the bladder

Umbilical fistula (Fig 120 (D)) —The umbilical end only remains patent, leaving a fistulous track running downwards This may become infected and discharge (*see* case B G , page 123) Lipiodol will indicate the direction and extent of the track

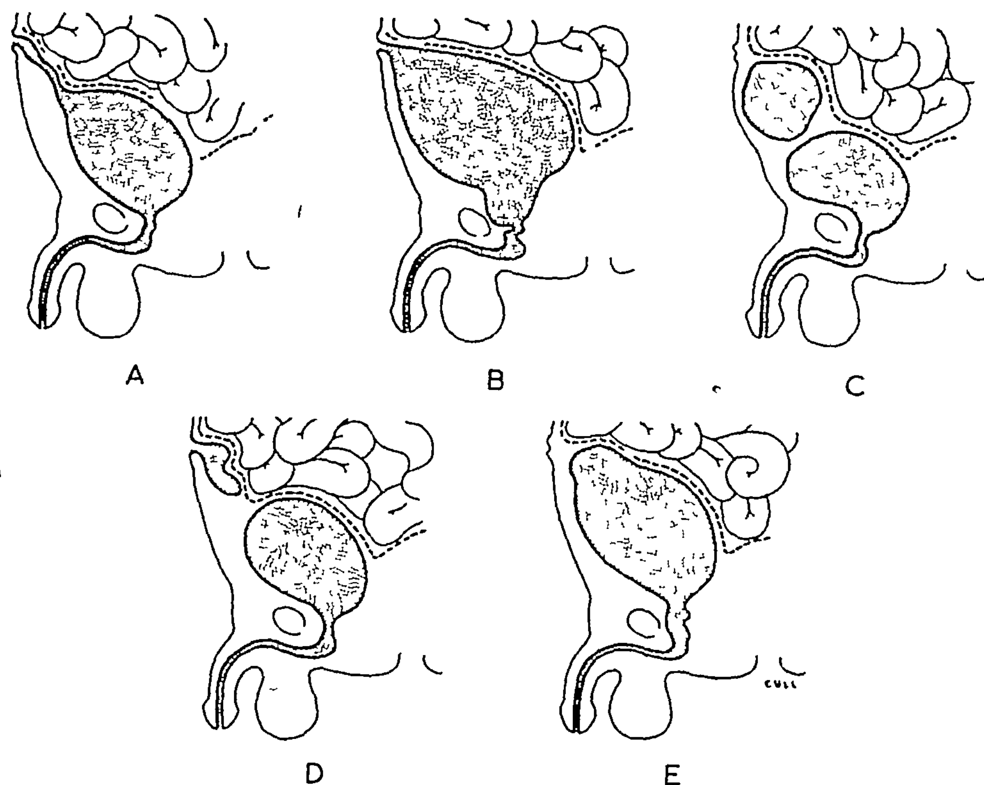


FIG 120 —Diagrammatic drawing to illustrate anomalies of the urachus
(A) Urachal fistula,
(B) umbilical fistula without formation of urachus,
(C) urachal cyst,
(D) blind umbilical fistula,
(E) urachal diverticulum at apex of bladder

Vesical anomaly of possible urachal origin —We have seen one case which suggested a rudimentary cap of urachal origin (urachal diverticulum) to the fundus of the bladder This was in a girl aged 10 years who suffered from a very persistent foul cystitis There appeared to be no obstruction to micturition and no involvement of the upper tract. The cystogram showed a large bladder with a persistent deformity of the fundus exactly as depicted in Fig 120 (E) Cystoscopy showed severe generalized cystitis with excessively marked trabeculation confined to the area of the fundus as outlined A partial cystectomy was performed with the aim

of removing this segment of the fundus The child was greatly benefited, though the urine is still mildly infected

Treatment

The treatment of urachal fistulae and cysts is by excision and it is usually not difficult to do this without opening the peritoneum An adequate mid-line incision is made and the track isolated by gentle dissection from the peritoneum Com munication with the bladder if present, is usually narrow and its closure presents no difficulty Drainage of the bladder is rarely necessary

RARE ABNORMALITIES

A brief reference must be made to the following·

Congenital urethral fistulae—These are seen on the ventral aspect of the penis, apparently resulting from localized defective closure of the urethral folds The main urethral channel is intact and micturition occurs in the normal manner, but there is of course a leak from the fistula. Such fistulae are apt to be trouble some and hard to close Where the opening is in the region of the coronary sulcus resembling the defect which sometimes results from a misplaced stitch during circumcision it is simpler to perform an extended meatotomy leaving the child with a glandal hypospadias

Diverticula of the anterior urethra—These have already been mentioned as a possible cause of obstruction to the lower urinary tract (*see* Fig. 52, page 116)

Diphallus—This is a rare teratological abnormality in which the child is born with an apparently double penis a urethral canal is commonly present in only one of the two organs, and where the child has been otherwise normal good results have followed amputation of the supernumerary penis

Pre penile scrotum—A case of this remarkable deformity has been reported by Huffman (1951) who found a similar instance in the literature Huffman s case died during the first month of life with urinary obstruction

Cloaca—A wide variety of anomalies involving persistence of a cloaca is reported in the literature In the majority of cases the infant is stillborn and no question of treatment arises Cases have, however survived and obviously present a very difficult problem in the solution of which it is not possible to lay down any rules for guidance

CHAPTER 15

DISEASES OF THE EXTERNAL GENITALIA

The prepuce and circumcision

Terminology

"PHIMOSIS" (Φιμωσις, muzzling) is defined as a "stenosis of the preputial orifice"

Acquired phimosis results from trauma If the preputial ring is so forcibly stretched that cracks appear, fibrotic contracture is very likely to follow A badly planned and ill-executed circumcision frequently ends in a contracted and fixed cicatrix, that is, an acquired phimosis

In the elderly, the same contracture follows from loss of elasticity or infection due to dirt and neglect The phimosis associated with carcinoma of the glans commonly has such an aetiology, and should not be confused with so-called phimosis of infancy, sometimes called congenital

So far as the child is concerned, it would be well if the term "phimosis" were confined to the acquired lesion Unfortunately tradition applies it loosely to the infant's normal prepuce, leading thereby to the firmly rooted conception of a pathology which is in fact non-existent

Embryology

The normal development of the prepuce, together with the development of the urethra, is described in Chapter 6, but it may be repeated here that the preputial sac is formed by the splitting of an originally solid lamella of cells, this splitting does not start until late in foetal life and is not necessarily complete at birth

The normal prepuce

In every new-born boy the prepuce completely ensheathes the glans "like a glove" and is partially fused to it by embryonic adhesions The extent of these and therefore of preputial mobility has considerable variation within normal limits The natural evolution is for the adhesions gradually to disappear, and for the prepuce to become mobile and retractable in conformity with the growth of the penis The process takes some months and it is quite erroneous to expect the infant's prepuce to retract anything like completely for some considerable time after birth (up to 9 months or longer)

Function

This being the natural arrangement, we may ask ourselves whether the ensheathing prepuce subserves any functional need Protection of the underlying glans from injury is obviously suggested That there is such a source of potential injury in ammoniacal dermatitis will be manifest when we come to consider this condition It should therefore be appreciated that the infant's prepuce is a normal anatomical structure designed for an important physiological purpose Indications for its removal must therefore be convincing and cogent The traditional "indications" commonly given for circumcising infants are

(1) The "tight" prepuce is alleged to be a cause of straining and to produce obstruction to micturition and even to lead to serious back pressure effects in the urinary tract. There is no evidence to support these views In infants dying of the back pressure effects of some such lesion as urethral valves, the cause has no doubt in the past been wrongly attributed to so-called "phimosis" which of course was clearly present The supposed association is in fact part of the general misconception and no children's urologist of experience would accept the "tight" prepuce as a cause of urinary obstruction Very occasionally where the preputial orifice is very small and becomes temporarily "gummed up" micturition results in momentary "ballooning" of the prepuce but such cases are never associated with distension of the bladder or any real obstruction to micturition The supposed effects of "straining" upon hernia rectal prolapse and the like are correspondingly mythical

(2) Cleanliness— balanitis Inability to retract the infant's prepuce is alleged to lead to lack of cleanliness and what is quite wrongly called "balanitis" The term balanitis should be confined to infection and suppuration occurring in the preputio-glandal sulcus This is only seen occasionally in older boys and in them is certainly an indication for circumcision What is often called balanitis in the infant is the condition illustrated in Fig 121 (*a*) This is an oedema of the tip of the prepuce seen very commonly in association with ammoniacal dermatitis Far from being an indication for circumcision this condition is in fact the strongest possible contra indication If in such a case the prepuce is removed the underlying glans is no longer protected and ammoniacal burning and meatal ulceration is virtually certain to follow To sum up the idea that the normal non retractile prepuce of the young infant is a potential source of infection is entirely without foundation

(3) The operation is advised to eliminate supposed reflex effects for example micturition disorders masturbation and so on Such "indications" are merely empirical without any pathological basis

(4) Circumcision of the infant is commended as a wise precautionary measure to prevent carcinoma of the penis 60 years or so later in his life, or even of the cervix of his prospective wife! This does not seem a very weighty consideration particularly if the common aetiology of the phimosis seen in old men under such circumstances is appreciated

(5) The operation is advised as a prophylactic against venereal disease later on No one questions that a non retractable prepuce is undesirable in an adult, whether he be a chaste husband or a moral weakling, but this affords no adequate reason for assuming that an infant's normal prepuce will not continue its natural evolution still less for depriving him of an appendage designed for his welfare

(6) Having the operation done early The argument runs that if a child is going to require circumcision ultimately it is naturally desired particularly by the parents to get the operation done early If it is clear that real need for the operation will arise ultimately in only a small proportion of children it is surely unreasonable to subject every infant to the procedure on the off-chance that he may prove to be one of the less fortunate few

There is in fact no convincing reason on surgical grounds for circumcision in early infancy, and many infants would be saved much misery if the practice were abandoned, the baby left in peace, and the parts cleaned in the ordinary way at the daily bath, a little white vaseline being gently applied to the penis regularly It will be found in the vast majority of babies that the prepuce retracts freely and easily after a few months If after 6–12 months the process appears too dilatory, particularly if there appears to be any real risk of paraphimosis or in the rare instance of "ballooning", then the baby should be anaesthetized, any residual preputial adhesions gently separated with a probe and, if need be, the preputial orifice sufficiently enlarged by making an adequate dorsal slit with scissors

In older boys a non-retractable prepuce may certainly give rise to complications, balanitis with purulent discharge, paraphimosis and even possibly enuresis from drag of the prepuce upon the external urinary meatus during erection These are undoubted indications for circumcision, but it should be realized that such persistence of non-retractability is a comparatively uncommon event and most boys escape these complications if left alone.

Ammoniacal dermatitis

A correct appreciation of the nature and aetiology of ammoniacal dermatitis ("napkin rash") is essential to prophylaxis and treatment

Hamilton and Middleton (1927), recording their experiences in Edinburgh, gave a masterly account of the whole subject, for which grateful acknowledgment must be made

Clinical features

These are well known They comprise the rash with which goes, in the circumcised infant, meatitis and ulceration, dysuria and micturition disturbances, and ammoniacal-smelling napkins

The rash is seen in the napkin area as a red, angry erythema with characteristic glazed patches, oedema and desquamation, secondary infection may intensify the lesion and even lead to serious ulceration In the uncircumcised boy the tip of the prepuce becomes oedematous and swollen (*see* Fig 121 (*a*)) In the circumcised boy blisters and ulcerated patches may appear on the glans and the meatus be reddened and everted or the seat of an "ulcer" The "meatal ulcer" causes the child great agony. The ulcer scabs over and makes micturition difficult or impossible to start When with much straining micturition is finally started, the tearing open of the crack causes severe pain, with screaming, reflex stoppage of the stream and, not infrequently, the escape of a drop or two of bright blood, which naturally adds to the general alarm and distress. Acute retention is a commonly associated feature

In the girl the urethra is protected by its position and the manifestations are less distressing in consequence

The napkins smell strongly of ammonia and "make the mother's eyes run".

Relation to feeding

The condition is virtually not seen in the breast-fed baby until weaning and is clearly related to artificial feeding (*see* below)

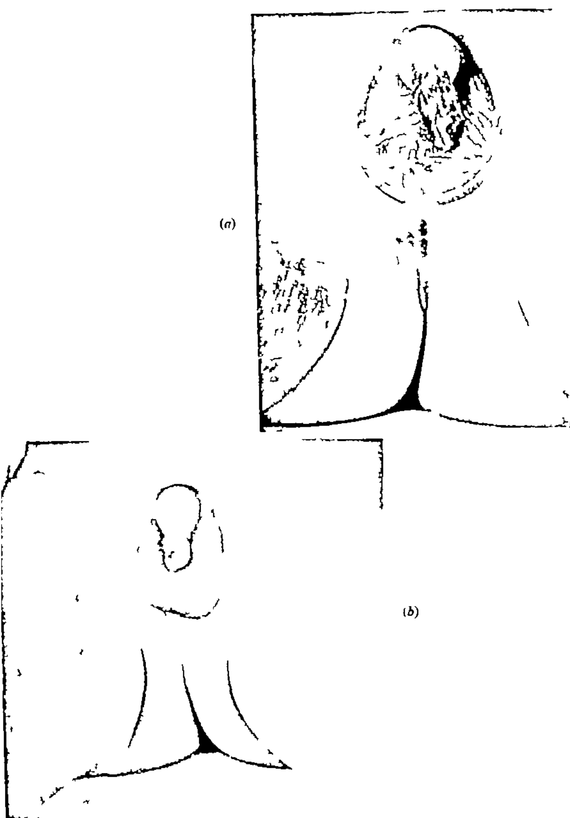


FIG 121 —Ammoniacal dermatitis.
(a) With severe involvement of the prepuce (b) the same after treatment

Explanation of ammoniacal dermatitis

The lesion was first established as a clinical entity by Zahorsky (1915) and by Brennemann (1921) and Cooke (1921) It was recognized that the condition was in fact an ammonia burn Cooke's researches showed that the source of ammonia was the urea of the urine, liberated by the action of an intracellular ferment (urease) present in a bacillus commonly found in the colon of children Cooke called this organism the *B. ammoniagenes* and described its characters

The organism is gram-positive, non-motile and aerobic It flourishes in an alkaline medium When inoculated into fresh sterile urine, ammonia is rapidly produced Cooke produced the typical skin lesions experimentally on his own forearm

The lesions, as seen clinically in the child, are therefore caused by ammonia liberated from the urine reacting with the bacillus in the confined atmosphere of the soaked napkin

In the breast-fed infant, the stools maintain an acid reaction, the bacillus does not thrive and the fermentation is unlikely to occur, but bottle feeding changes the reaction of the stools to alkaline, thus activating the fermentation process

In children past the infant stage a similar change of reaction may follow dietetic disturbances accounting for periodic "ammoniacal storms" (Hamilton and Middleton, 1927), which may determine meatal ulceration in little boys of 2–3 years

Prevention and treatment

Here then is a condition of such clear aetiology that its prevention and treatment are eminently simple

(1) If the infant were always dry and breast-fed there would be no ammonia burn, and there is therefore another significant urge to maintain breast feeding, as well as to encourage micturition training and dry napkins

(2) Since the *B ammoniagenes* will not thrive in an acid medium, the liberal use of boracic acid will check the fermentation This is achieved by careful removal of all traces of soap (alkaline) from the napkins by frequent rinsings after washing, and by impregnating the napkins with boracic acid powder

(3) Since the burns are only likely to occur when ammonia vapour acts in a closed space, the child when sleeping, and likely to be wet for long periods, should not wear napkins at all The clothing should be loose and the requisite towelling and mackintosh be spread out under him

(4) It is sometimes attempted to change the reaction of the urine by diet, but this seems to be of little value Small doses of acid sodium phosphate may be helpful in reducing the *p*H of the urine, and if secondary infection has occurred local applications of penicillin sulphathiazole cream may be used.

Meatal ulcer

The preventive treatment of this lesion is the avoidance of circumcision. It is practically never seen in the uncircumcised infant The treatment of the lesion when it occurs is mainly that of the associated ammoniacal dermatitis When the ulcer is covered by a scab making micturition difficult, the baby is greatly helped by softening the scab in a bath or by sponging Local applications of boracic acid ointment are necessary, and in older boys protection from friction by the trousers.

The view is often expressed that the meatus is congenitally small and that this is an important aetiological factor in the ulceration We do not believe this is often the case, and in our experience meatotomy is seldom called for Contracture of the orifice may result occasionally from scarring and the operation will then be advisable. No decision as to the need for meatotomy should be taken until some months have elapsed after the healing of the ulcer and the operation should never be done until all risk of re ulceration is past

Other infections of the genitalia

Occasionally involvement of the urethral meatus and adjacent skin as part of a generalized cutaneous infection such as impetigo or eczema may lead to retention of urine.

Generalized vaccinia is one such condition but occasionally the secondary spread of the vaccinia actually involves the genitalia from implantation by the child s fingers the local inflammatory change can be very severe

There are however other conditions which occur both in adults and children, and have a particular significance in that the genital lesion is a specific part of the general condition. These infections fall into two groups the aetiology and associations of which have not been clearly elucidated.

The first of these, which is now commonly referred to as erythema exudativum multiforme, covers a group of conditions in which there is a febrile illness with surface lesions affecting the skin around the mouth, the buccal mucous membrane and the genital skin and the conjunctiva There are sometimes also diffuse cutaneous eruptions which may be papular or bullous. The more severe type associated with prolonged fever has been commonly referred to as the Stevens Johnson syndrome although this is not historically correct, and the milder form is described as the Hebra type (Thomas, 1950 Shaffer and Morris, 1948) The association of ophthalmia with the skin and genital lesions and fever is a fairly common finding.

The second group of conditions is known as Reiter s disease in which there is urethritis and conjunctivitis and arthritis usually involving the larger joints such as the knee The urethritis may present as meatal ulcer crusting and be the first symptom to call attention to the existence of the condition which might otherwise pass as a simple arthritis This condition has been recently reviewed by Corner (1950) in its particular relation to children Reiter s disease was first described in 1916 and there appear now to be two distinct forms—the venereal and the post-dysenteric The latter is the one which affects children and its appearance seems to be preceded often by an attack of dysentery with a Shigella Flexner infection. The treatment of these two conditions is still unsatisfactory some responding to the more recent antibiotics such as streptomycin and aureomycin and some failing to respond to any known drug. The infecting organism is most likely to be a virus

When faced with any such eruption the clinician must bear in mind the similarity between these muco-cutaneous-ocular syndromes and certain drug eruptions, especially from sulphonamides. Since such drugs are frequently used in the treatment of dysenteric infections, the confusion may be very difficult to avoid and the involvement of large joints in drug sensitivity reactions is also not uncommon.

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Urethral caruncle

Small globular nodules of a deep red or purple colour are occasionally seen on the urethral lip of little girls These are usually painless, though they may cause local irritation and aggravate vesical irritability Their histological structure is that of a haemangeioma with a varying fibrous stroma Similar lesions are not infrequently seen at the anal margin in both sexes. They are best destroyed by diathermy We have not met in childhood the acutely tender caruncle as seen in adults

Prolapse of urethral mucosa (Fig 122)

This must be distinguished from the foregoing Urethral mucosal prolapse has been seen by us on 3 occasions In all there was an associated urinary infection with frequency and straining The onset of the prolapse is comparatively sudden and the condition is very painful The protruding mucosal ring rapidly becomes oedematous, congested and inflamed

The treatment is conservative complete rest in bed with local detergent applications (lead and opium compresses) and general sedatives, together with appropriate attention to the urinary infection if still present The condition slowly resolves No sloughing occurred in our cases, the inflammation subsided in 2–3 weeks and the part returned to normal without recurrence

The temptation to interfere by fulguration or excision should be resisted

Vulval adhesions (Fig 123 (*a*) and (*b*))

It is comparatively common in little girls to see the labia minora fused together by a pearly grey continuous membrane often complete right up to the urethral meatus To those who are unfamiliar with the condition, the appearance suggests an atresia of the vagina and it is the fear of this that commonly brings these children to us

The condition is very simply dealt with by separating the adhesions with a probe The procedure causes the child a little momentary pain and an anaesthetic is usually desirable The separated edge bleeds slightly and careful cleansing and frequent smearing with sterile Vaseline or penicillin cream is necessary for a week or two The nature of these adhesions is doubtful They are probably congenital but an associated vulvitis is common and cannot be entirely ruled out as a causal factor.

CHAPTER 16

HERMAPHRODITISM AND DISORDERS OF SEXUAL DEVELOPMENT

Intersex

In the past the hermaphrodites have often been regarded as curiositie considerable theoretical interest but so rare as to be of little practical concern the urologist This viewpoint underestimates however the number of case which the form of the external genitalia occasions difficulty in the determina of sex, and the decision as to the upbringing of the child is not one to be lig undertaken This chapter is concerned with the practical difficulties encount in dealing with these cases, and a review of the theoretical implications man them unsolved problems is outside our scope

The attributes commonly recognized as sexual are (1) the gonads (2) secondary sexual organs. penis, scrotum prostate seminal vesicles, vas epididymis in the male clitoris vagina uterus and tubes in the female (3) general bodily form muscularity distribution of hair and many other feat more easily recognized than described and (4) the psychological orienta towards the opposite sex In childhood this may not be altogether evident, tho some indication can be gained from the habits of play and choice of playm It would be a mistake however to assume that normal boys never play dolls, or that mechanical aptitude is unknown amongst girls

It is a matter of common observation that normally developed gonads genitalia are by no means invariably accompanied by a full masculinity femininity of bodily form or psychology and the intersexes in whom the exte genitalia are not fully characteristic of either male or female must be regarde extreme examples of a common tendency It is important to realize that psychology the sexual behaviour or inclination does not necessarily corresp either to the type of gonad or to the form of the genitalia, and it is convenien refer to the pragmatic sex of an individual a term introduced by Cawadias (1 to indicate the sex in which the person feels himself or herself to be

Determination of sex

So far as can be judged at present, the sex of the embryo is determined at time of fertilization of the ovum by the genes upon the X and Y chromoso The reduction division which takes place during the maturation of the ga leaves the ovum with only an X chromosome whereas the sperm may pos either an X or a Y The combination XX determines a female XY a male emb and the sex thus determined is known as the genetic sex. The manner in which genes produce a male or female body is largely obscure in some animals, as in insects for instance local action of the genes appears to be entirely responsible the whole bodily form In the higher vertebrates, however it is certain that inte

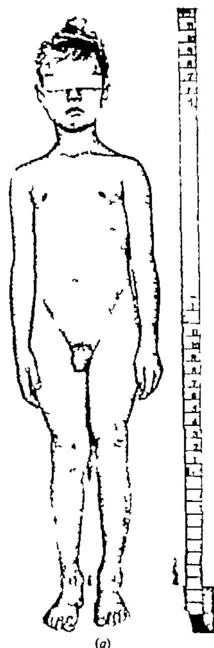
secretions play a large part in differentiation The embryo at first possesses the potentialities of either sex, and the primitive gonad contains elements both of testis and ovary. By the 13–15 millimetre stage the differentiation of the gonad is evident and at a somewhat later period the development of the genital passages becomes distinctive for either sex This latter development unquestionably proceeds under the influence of a secretion of the gonad, but whether in fact this secretion is the same as the hormone derived from the fully developed organ appears to be doubtful The adrenal glands are also of some importance in the development of sex, though their normal action is less well known than their pathological effects The adrenal cortex is derived from a mesothelial proliferation on the genital ridge (8 millimetres) dorsomedial to the gonadal area, and in foetal life is chiefly composed of a mass of acidophil cells These constitute the X zone or foetal cortex and undergo involution after birth, leaving the adult cortical cells to differentiate on their surface The foetal cortex has been said to contain an androgenic substance, but its exact role in normal development is obscure

It will be clear from the preceding discussion that deviations from the normal sexual development may occur at several stages and that later deviations will be more easily identified than earlier Thus if the gonad is normally formed but a hormonal disturbance distorts the pattern of subsequent development, it will still be possible, by finding a testicle or an ovary, to decide the true sex, and these cases are spoken of as "pseudohermaphrodites" Should the error occur earlier in embryonic existence, however, elements of both testicles and ovary may be present in the same individual, who may then be described as a "true hermaphrodite". This term is a convenient one but not strictly accurate, since we have no means of determining the genetic sex of an individual, and bisexual fertility (which is the characteristic feature of the animal hermaphrodites) does not occur in man

Female pseudohermaphroditism

In the female foetus, the form of the external genitalia may be distorted as a result of excessive output of androgenic hormone by a hypertrophied adrenal gland The ovaries are normal and under their influence the Mullerian ducts differentiate as vagina, uterus and tubes, while the Wolffian ducts degenerate The virilizing effect of the adrenal cortex then comes into operation, this prevents the normal disappearance of the urogenital sinus and at the same time stimulates the growth of the phallus and of the labio-scrotal folds The vaginal orifice, therefore, may or may not be evident to external inspection, depending on the degree of fusion of the urethral folds, but in all personally observed cases it has opened at the same level relative to the proximal urethra the level it has reached in a normal embryo of 80–100 millimetres CR length, which is already considerably below the situation of the definitive verumontanum in the male

In the least severe cases of this deformity, the vaginal orifice can be displayed by firm retraction of the labia, and although the clitoris is considerably enlarged these children are usually brought up according to their true sex This type is, however, not so common as the more advanced cases, in whom the adrenal stimulus has presumably started earlier and who have but a single external orifice the genitalia closely resemble the condition of perineal hypospadias in the male The most severe degree of this condition is seen when the labio-scrotal folds have



(a)



(b)

FIG 124—Female pseudohermaphrodite (Mr Denis Browne's case) The photograph (a) shows the appearance of the child at the age of 3 years and 10 months.

"He had been brought up as a boy and behaved as one. His growth was considerably advanced for his years, the phallus was large and erectile, and pubic hair was well developed. There was a penile hypospadiac urethral orifice. Bone age was 11 years and ketosteroid excretion 14 mg. per 24 hours. Urethroscopy was performed and the vaginal orifice identified in the floor of the urethra. The instrument was introduced into the vagina and the cervix catheterized (b) shows the salpingogram produced by the injection of pyelosil into this catheter The appearance confirmed the diagnosis of female pseudohermaphroditism, but in view of the masculine physique and psychology it was decided that the child should be brought up as boy (By courtesy of *Proceedings of Royal Society of Medicine*.)

fused in the mid line to form an empty scrotum and the urethral meatus is placed on the under surface of what appears to be a large penis The vaginal orifice may then be discovered by endoscopy as the instrument is withdrawn down the urethra it will be noted that the verumontanum is absent, although there may be a few short duct openings on the posterior urethral wall which presumably represent enlarged Skene's tubules The external urinary sphincter does not suddenly obliterate the field as in normal urethroscopy but at approximately the region of the perineal membrane the gaping orifice of the vagina may be found Polypoid mucosal tags may surround this orifice and obscure it the eye piece of the urethroscope must be raised almost to the vertical in order to search this region carefully In most cases it is possible to introduce the instrument into the vagina, visualize

the cervix and catheterize it Salpingograms will then confirm the diagnosis (*see* Fig 124)

The effects of the adrenal over-activity do not cease at birth, and precocity, with continuing virilization, may soon be evident Growth is rapid, ossification and epiphyseal fusion in advance of normal, muscular development, growth of pubic hair, and enlargement of the phallus are increasingly prominent Ovarian function is normally suppressed and menstruation absent, but one case in which menstrual bleeding appeared as haematuria and caused clot retention has been recorded (Everidge, 1945) Prostatic tubules may develop around the proximal urethra The androgenic hormone is excreted in the urine as 17-ketosteroids in excessive amounts The psychological development of these children is chiefly upon masculine lines, and despite their genetic femininity they are often pragmatic males Exceptions to this rule undoubtedly occur, however, and it is unwise to be too dogmatic in prognosis or too hasty in undertaking the surgical correction of the genital deformity

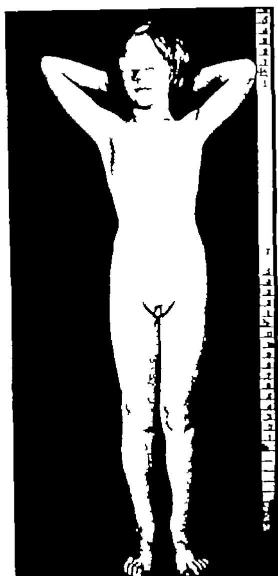
Occasionally the excessive androgen output of the adrenal is accompanied by a deficiency of the other endocrine functions of the gland, and the infants, in addition to their pseudohermaphroditism, have the signs of Addison's disease. Such cases may present clinically on account of anorexia, weakness and vomiting Crises occur with diminution of the serum sodium and corresponding elevation of the potassium (Panos, 1950).

Male pseudohermaphroditism (*see* Figs 125 and 126)

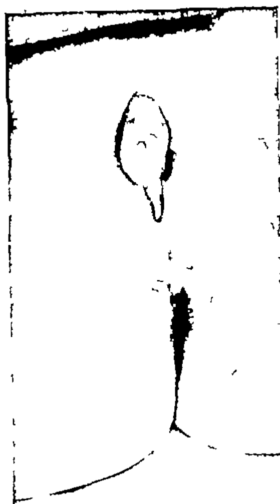
Male cases of pseudohermaphroditism do not follow any such characteristic pattern as has been described in the female, and no hormonal cause has yet been established Difficulty in the determination of sex may arise from a simple failure of the normal development, and it is hard to define the exact point at which hypospadias is of such a degree that the term pseudohermaphroditism is appropriate Confusion of sex has frequently arisen in children with perineal hypospadias, a small penis and undescended testicles, but in addition cases are occasionally encountered in which the male has a small blind-ending vaginal pouch, sometimes evident on parting the labio-scrotal folds, sometimes concealed within the urogenital sinus as in the female pseudohermaphrodites We have observed a case of a male with complete, though undeveloped, vagina, uterus and tubes, and such individuals present a formidable problem in diagnosis Such perversions presumably result from a genetic error which has altered the reaction of the somatic tissues to the normal gonadal hormones, since endocrine abnormalities have not been observed in these cases, and adrenal hypertrophy during the foetal life of a genetic male results in precocious development along isosexual lines, not in feminization The pragmatic sex of the individual is usually male, but pragmatic females have been described by Novak (1935) and by Young (1937) It is difficult to know to what extent the sexual attitude of the child is determined by his make-up and to what extent by his upbringing

True hermaphroditism

Individuals possessing both testicular and ovarian tissue are very rare, and a diagnosis of true hermaphroditism can only be established by histological examination of the gonads All cases reported up to 1937, in which adequate



(a)

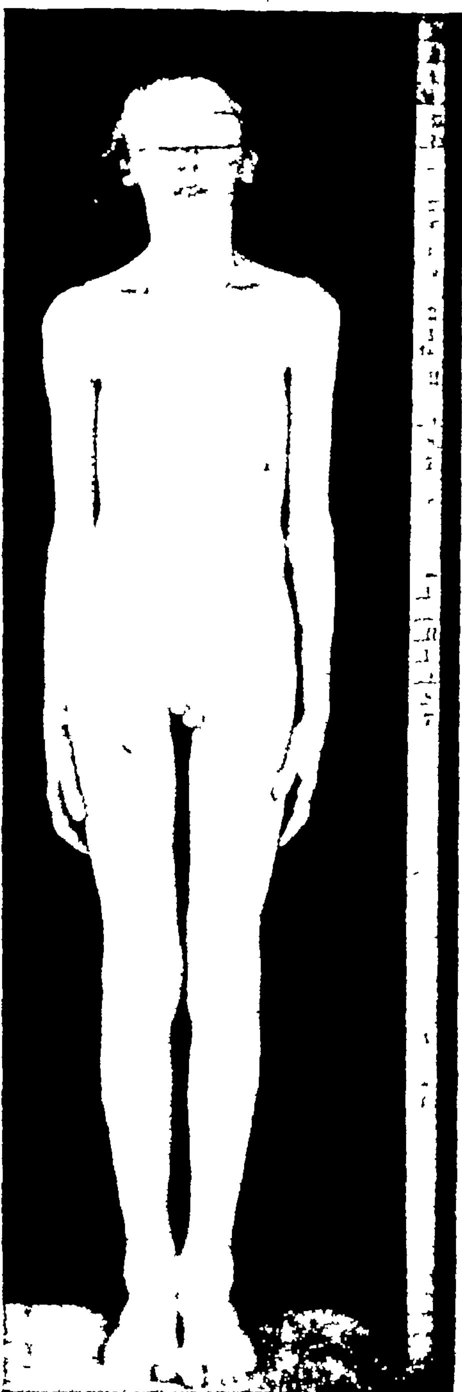


(b)

FIG 125—Male pseudo-hermaphrodite. Aged 8 years.

The patient was brought up as girl but found to have a small well-formed penis with a perineal hypospadias and marked chordee. There was very little evidence of a scrotum but both testicles were palpable in the inguinal region. A well-formed vaginal pouch was present behind the urethra. Ketosteroids were 1 4 mg per 24 hours A laparotomy was performed and no female organs found. The hypospadias wa repaired by the Denis Browne method, leaving the vaginal pouch untouched, and the child was very much happier in the male role.

details were available have been reviewed by Young. The great majority had not presented themselves until after puberty when urethral menstruation or the development of the breasts in a child believed to be male had drawn attention to the anomaly Many have been diagnosed by the discovery of an ovary in a hernial sac No rule can be laid down regarding the form of the genitalia most cases have had a hypospadiac type of penis and in addition a vagina opening exteriorly or into the urethra There may be a testicle on one side and an ovary on the other or the gonads may contain areas of tissue characteristic of both (the ovo-testis) The pragmatic sex may be ill-defined but is more often entirely male or entirely female



(*a*)



(*b*)

FIG 126—Male pseudo-hermaphrodite Aged 12½ years

The child had been brought up as a girl and was said to behave as one She was noticed to have a swelling in the labia and was brought for examination There was a dwarfed hypospadiac penis with a perineal urethral meatus and a split scrotum The left testicle was fully descended, the right in the external inguinal ring. Nothing was to be felt P R The 17-ketosteroids were 3 3 mg per 24 hours A diagnosis of male pseudohermaphroditism was evident *He was referred elsewhere for plastic operations to* improve his appearance

Diagnosis

It is of vital importance to realize that the diagnosis of sex frequently entails a thorough investigation, and no treatment, either medical or surgical, should be undertaken before the full facts of the case have been established The permission of the parents for this investigation is sometimes difficult to obtain since the sex has normally been pronounced at birth, with surprising confidence, by the midwife, and a change will involve the whole family in acute embarrassment The

child is brought up with a view to plastic surgery which will fashion the genitalia according to its accepted sex.

Clinical examination should take into account both the bodily form and the mentality of the child, and it is important that the examiner should not be prejudiced either by the clothes or the hair-cut Evidence of precocity must be carefully sought and an estimate of the bone age by radiological examination of the epiphyses may give valuable information Measurement must be made of the 24-hour excretion of 17-ketosteroids

Examination of the genitalia is best performed under anaesthetic not only does this save the child considerable discomfort and embarrassment but relaxation is essential for adequate palpation and for endoscopy Where both testicles can be palpated the diagnosis may be at once evident, but it should be remembered that an ovary may well present itself at the inguinal ring and even in the scrotum dragging with it the fallopian tube If a gonad is felt, therefore particularly if it is unilateral the vas and epididymis must also be sought and should doubts still remain a biopsy is a very simple matter

The glans should be examined with care In some cases it can be seen that the groove which normally marks the ventral surface of the hypospadiac penis is absent and the glans has the normal rounded off appearance of the clitoris Such an appearance is very suggestive of a female origin but the presence of a groove cannot be accepted as a certificate of masculinity

The vaginal orifice must be carefully sought. Where the urethral meatus is in the perineum the vagina may be found by firm retraction of the meatal margins but otherwise the urethroscope should be employed and the posterior urethral wall carefully inspected The vagina or cervix may be catheterized and demonstrated in skiagrams after the injection of opaque fluid

Bimanual pelvic examination may establish the presence of a uterus but it is not possible to distinguish by this means between ovaries and intra abdominal testicles

If at the end of an examination under anaesthesia it can be said that the uterus and vagina are unquestionably absent, a diagnosis of masculinity can be made with some confidence but the converse does not hold and a vagina may be present in the male Where an occult vaginal orifice is accompanied by precocity and by excessive output of 17-ketosteroids the diagnosis is certainly female pseudohermaphroditism In the absence of endocrine disturbance, if the genitalia give no certain clue a laparotomy should be performed without hesitation and a biopsy taken from the gonads. Indeed this measure may be advised in any doubtful case It is only in this way that a true hermaphrodite can be identified.

Treatment

The guiding principle in the treatment of all this group of cases must be the adaptation of the external appearance to the psychology of the child rather than to the histology of the gonad For the most part this treatment will consist in the correction of hypospadias and in the removal of the superfluous female genital organs

It might be supposed that the female pseudohermaphrodite of adrenal origin could be treated along more fundamental lines and the endocrine balance

restored While this may well be possible in the future (and it has already been shown that the administration of cortisone causes a reduction in the output of androgens) it must be admitted that so far attempts at this restoration have been unsuccessful Partial resection of the hypertrophied adrenals is not followed by any permanent change, and the regular administration of oestrogen has had disappointing results In the female pseudohermaphrodite, therefore, an assessment must be made of the degree of virilization, most cases are happier as boys and should be allowed to continue as such The urethra should be reconstructed by the method described in Chapter 14, and where the uterus and vagina are well developed they should be removed In the mild cases, already brought up as girls and content to remain in that role, plastic enlargement of the vagina and amputation of the clitoris may be required

The male pseudohermaphrodites are usually happier in their true sex, and where they have hitherto been regarded as girls a change of upbringing may well be advisable in addition to plastic surgery to the genitalia It is unnecessary to remove a blind vaginal pouch

The true hermaphrodites are unpredictable and each must be treated on its own merits In cases brought up as males hysterectomy may be necessary to prevent recurring haemorrhage, and where ovarian tissue is separate from the testicular the former may be removed Interference with the gonads should be cautious, however, for if all the functioning tissue is removed or damaged, eunuchoid changes may result

Sexual precocity

The onset of normal puberty is apparently determined by an increased output of gonadotropic hormones from the pituitary In the male the spermatic tubules develop rapidly under the influence of this hormone and the testicles enlarge, subsequently enlargement of the external genitalia, growth of sexual hair, deepening of the voice, and other changes characteristic of puberty follow as a result of stimulation by androgens secreted both by testicles and adrenal cortex Androgen excretion, as measured by the output of 17-ketosteroids, rises In the female ovarian development occurs following upon the pituitary stimulus, but the external changes of puberty result partly from oestrogenic influence and partly from the androgenic influence of the adrenal cortex (*see* Wilkins, 1948)

True precocious puberty, as opposed to premature development of the external genitalia, must therefore result from disorders which act through the pituitary In fact the majority of cases show no other signs of ill-health, and ultimately develop into normal adults, this type is usually referred to as "constitutional" precocious puberty Intracranial lesions may be responsible, *via* the pituitary, for a similar premature appearance of sexual function

The urologist, however, is chiefly concerned with those rare cases in which growth of the external genitalia and development of the bodily form characteristic of puberty has preceded the pubertal change in the gonad. In boys this type of precocity may result from adrenal cortical tumour or hyperplasia, or from an interstitial cell tumour of the testicles Adrenal cases are discussed in Chapter 13, the testicular tumours with endocrine disturbance are very uncommon and often unobtrusive, but should not be missed if a careful examination is made The

androgenic excretion is considerably raised while the contra lateral testicle remains infantile in type. In the female the granulosa-cell or thecal-cell tumours of the ovary may be responsible for isosexual precocity but adrenal tumours always result in virilism. Jolly (1951) has shown that in all reported cases of ovarian tumour causing precocity the tumour was sufficiently large to be easily palpable so that in suspected cases a bimanual pelvic examination under anaesthesia is usually sufficient to exclude this very rare disease.

Microgenitosomia

Small boys are not infrequently brought up to the urologist because the penis is thought to be unnaturally small. In infants this appearance is almost always due to the fact that the true dimensions of the organ are obscured by the excessive development of the pad of fat over the symphysis. In older children the variation in the normal development of the genitalia is very considerable and caution should be exercised in the diagnosis of an abnormality but a true microgenitosomia is occasionally encountered either with undescended or with normally placed but undeveloped testicles. The testicular defect in these cases cannot be repaired although the size of the penis and the outward appearance may be improved by testosterone treatment. In adolescence the testicular agenesis may be associated with eunuchoidism and excessive development of the breasts.

Under-development of the female genitalia is scarcely ever noticed in childhood though an occasional case may be recognized during the investigation for enuresis.

We are glad to acknowledge the help we have received from Dr H Jolly in the elucidation of the problems of sexual development.

APPENDIX I

VESICO-URETERAL REFLUX

VESICO-URETERAL reflux figures so prominently in a variety of lesions that it is convenient to review here its nature and incidence in childhood

In the normal resting bladder the pressure is maintained at such a low level that the force of the ureteric contraction is sufficient to eject a brisk jet of urine into it After the efflux, the ureteric lumen is obliterated by the normal tone of its muscles. In contracting, the bladder muscle is capable of producing within it a pressure far above anything which the ureteric muscle is capable of withstanding, it is therefore necessary that there should be some mechanism to protect the upper urinary tract against regurgitation from below The obliquity with which the ureter traverses the bladder wall is undoubtedly a part of this mechanism, but the valvular effect is clearly greater when the bladder is distended, and in the empty bladder the ureter must be protected by the contraction of the muscles which surround its orifice

Vesico-ureteral reflux is more easily obtained in the experimental animal than in man, and more easily in some animals than in others perhaps the length of the intramural ureter is the important factor (Prather, 1944) Graves and Davidoff (1923) have given an admirable description of experimental reflux in rabbits Peristaltic waves normally pass down the ureter at intervals of 1–5 minutes and are normally of sufficient strength to obliterate the lumen at the point of contraction, the ureteric orifice opens only as the wave of contraction reaches it If the bladder is now progressively distended, the force required to drive the column of fluid into the bladder steadily rises, and after a time the ureteric musculature becomes unequal to the task The lower end dilates, the waves of contraction now no longer obliterate the lumen so that, as the orifice opens, the fluid in the bladder, under a high pressure, is able for a moment to flow up into the whole length of the relaxed ureter, unless, as sometimes happens, fresh and more powerful waves appear and push it back again Anti-peristalsis, although an occasional possibility, plays no part in this phenomenon In man a simple raising of the pressure in the bladder seldom produces reflux, the valvular mechanism being more effective than in the rabbit However, when cystograms are employed routinely in the investigation of minor conditions such as enuresis, reflux will occasionally be observed, particularly if the filling of the bladder has provoked a powerful contraction In such cases the reflux does not usually fill more than the lower half of the ureter with dye The valvular mechanism is more easily disturbed in children than in adults and reflux occurs in a variety of conditions

Congenital (The "ear-trumpet" ureteric orifice) —In this condition, which is described in the chapter on megaureter, we believe that the musculature around

the orifice has not been adequately formed and fluids introduced into the bladder flow freely into the upper urinary tract even under low pressure (*see* page 149)

Inflammatory—A severe cystitis may cause such oedema in the neighbourhood of the ureteric orifice that the tissues are rendered rigid The lumen can then be obliterated neither by the flattening which normally occurs on distension nor by the muscular contraction It has been shown experimentally in dogs (Auer and Seager 1937) that if the neighbourhood of the ureteric orifice is made oedematous by the injection of magnesium sulphate reflux occurs more readily and at lower intravesical pressure than in controls Moreover the effect on the bladder muscle of the cystitis is to make contractions stronger and more frequent.

Obstructive—Although acute and rapid rises of pressure in the uninflamed bladder seldom cause reflux chronic retention in children is not infrequently the cause of this phenomenon It appears that in time the valve gives way though it may be noted that in cases of lower urinary obstruction in which there is extreme hypertrophy of the bladder the hypertrophied detrusor itself may constitute a bar to reflux, and a greatly dilated ureter may still not fill from below It has been observed on a number of occasions that in infants with infra vesical obstructions there may be a reflux at the first examination made soon after the acute retention but after a period of catheter drainage, reflux can no longer be demonstrated. The ureteric muscle which gave way has had time to recover its tone

Mechanical dilatation of the orifice—Immediately after the passage of a large stone from above, or a large bougie from below the ureteric orifice allows reflux Persistence of this condition is unusual and recovery is rapid in the absence of inflammation Ureteric meatotomy needs to be very complete and to lay open the entire length of the intramural ureter to produce reflux even after the excision of a ureterocele reflux is by no means invariable

Neurogenic—In the neurogenic bladder due to spina bifida, the common lesion in children reflux is frequent It is almost a constant feature where there is any dilatation of the upper urinary tract. Experimentally (*see* Jacobson 1945) reflux occurs with regularity in the autonomous bladder (for example cauda equina lesions) less often in the automatic bladder resulting from high cord section and in the atonic bladder resulting from section of the afferent nerve roots It is reasonable to attribute the reflux to the comparatively high pressures which are maintained by the neurogenic bladder over a long period of time and to the irregularity of the detrusor contraction which perhaps throws out of action the valvular effect. Section of the ureteric innervation alone has no effect either upon the normal ureteric contractions or upon reflux (Wharton 1933 Barksdale and Baker 1930)

Although the vesical muscle is clearly very much more powerful than the ureteric muscle, a simple reflux from a normal bladder is not sufficient to produce permanent dilatation of the ureter Vermooten and Neuswanger (1934) observed dogs in which the uretero-vesical valve had been completely destroyed by section reflux only produced dilatation in cases in which the urine was infected This

observation is in accordance with clinical evidence Not only does the inflammation provoke frequent bladder contractions but it has an effect upon the ureter analogous to paralytic ileus

It is clear therefore that many factors may be concerned in the production of ureteral reflux in childhood, and that the extent of the ureteral dilatation and the rapidity of its development varies greatly in different circumstances Gross dilatation of the ureter is not in itself necessarily disastrous It would seem in fact to be an important protective adaptation Limitation of the dilatation at the pelvi-ureteric level is often impressive, so that a comparatively small and normal looking renal pelvis is seen surmounting a huge ureter, which by its very size is effectively buffering the kidney ("defence in depth")

APPENDIX II

TECHNIQUE OF CYSTOMETRY IN THE CHILD

CYSTOMETRY is not an exact investigation in the sense that may be applied to a chemical analysis or a radiographic visualization and we would emphasize that it is a procedure which should never be delegated to someone who is inexperienced and unfamiliar with its use any more than should cystoscopy The method which is described has been found to be suited to use in children and the interpretation of findings depends upon familiarity with the method which is soon acquired

The method recommended is to use a simple continuous gravity flow supply fed through the open top of a vertical manometer Other methods have been devised including those which depend on the intermittent delivery of measured quantities to the bladder with subsequent serial pressure readings, but this may involve, particularly in children the stimulation of bladder muscle by the rapid volume increase We favour the continuous flow method as more nearly reproducing the normal filling effect although the procedure takes longer

The simplicity of the apparatus eliminates unnecessary tubing and the use of a double column of fluid, which is otherwise present if the flow is fed in at the bottom of the manometer The glass tube and scale is that supplied with the standard Riches-Dukes tidal drainage apparatus A special flask container is required of a diameter of 2 inches or less in order that the relatively small 20 millilitres increments may be measured with reasonable accuracy It may be necessary to graduate the flask A rubber catheter is used No 4 or 5 F in children under 6 years of age but the largest which can be introduced easily is the best. All catheters should be carefully inspected as in many of the smaller rubber ones there is a marked constriction of the lumen just proximal to the eye which would be sufficient to restrict the ebb and flow of the fluid, and would therefore invalidate the record Such a stricture needs to be reamed out.

Cystometry needs to be carried out with as little fuss as possible and in an environment which will not scare or startle the child. The entry of a nurse, the dropping of a bowl the banging of a door or unnecessary small talk on the part of anyone present may produce abnormal bladder contractions, and will in any case interfere with the co-operation of the child

All but a very small number of children are tolerant, co-operative and excellent as witnesses The purpose and procedure should be explained to the child after the insertion of the catheter and the apparatus should be placed so that the child can see the manometer and co-operate in maintaining a low and steady pressure

The fluid used is fresh warm potassium permanganate 1 10 000 in water Excessive heating produces a brown precipitate in the solution Air must be expelled from the apparatus and sufficient fluid run through to warm the tubing.

Immediately before the insertion of the catheter the child voids urine naturally and then the residual urine, if any can be measured on the same occasion A

catheter specimen can also be collected for bacteriological examination If cystography can be carried out at the end of this investigation, one catheterization will have served all the necessary tests, and the child will be spared the inconvenience of repeated instrumentation We have on numerous occasions, by the introduction of local anaesthetic solution during the withdrawal of the catheter, been able to cystoscope both boys and girls as young as 5 years, but the ease with which this may be done depends largely on the size of the meatus, which cannot be adequately protected against the pain of stretching by topical application

The manometer is adjusted so that the zero mark is level with the symphysis pubis The fluid is then run in slowly, at first approximately at 60 drops a minute, but when 40–60 millilitres have been introduced the rate of drip can be increased. The rate should never become a continuous stream, and the operator will very soon be able to tell the rate at which the bladder muscle will relax in any individual There should in normal older children and adults be a rise of less than 2 centimetres pressure for each unit of 20 millilitres introduced If the rise is more than this at the beginning, it is more than likely that the drip rate is too fast

The normal investigation will reveal a gradual rise of base pressure to approximately 20 centimetres, this point being reached at volumes which vary mainly according to the age and size of the patient The total tolerated volume, though important, is of far less significance than the pressure or the presence of systoles

Usually between the second reading (40 millilitres) and the point of imperative micturition ("end-point") there are 2 normal desires to void (N D V) These are sharply defined points and the child should be forewarned and asked to announce their arrival He is each time told to take some deep breaths, and usually the sensation rapidly passes off* Systole should not normally occur at these points, but if it does the bladder will again relax rapidly as the child breathes deeply The maximum pressure is recorded for each contraction and deep deliberate breathing is continued until diastole is complete (to the "pre-systolic base pressure")

The filling procedure is continued until the child is "unable to wait any longer" The end-point is invariably heralded by flexion movements of toes, and often inversion of the feet, circum-oral pallor and flushing of the cheeks If a terminal rise of pressure occurs, its height should be recorded and the drainage clamp released We do not think that observations on pressures during emptying give any useful information *Care must be taken not to overdistend a bladder which has been rendered insensitive by nerve damage or infection*

Findings

At the end of the investigation, which may last 20–30 minutes, most children are composed, and one has even fallen asleep The following information should then be available (1) the pressure gradient during filling, (2) normal desire to void and the points at which this has occurred ("D" or "N D V "), (3) transient systoles (perceived) occurring with the desire to void the pressure reached is noted and recorded as "D 33" for instance, (4) the end-point or point of imperative micturition with the pressure of a terminal systole if it occurs ("I M " or "I M 44") (The end-point may come actually after the commencement of a contraction of

*These uninhibited contractions may often be seen to be bi-phasic, the first rather slow rise does not reach consciousness, there follows a very brief and partial relaxation and then a sharper greater pressure rise which is perceived by the patient

which the early phase was not perceived Such is a common finding in the enuretic child with day urgency or day wetting.) and (5) filling sensation

In preparation for recording on a graph, the observer may make his notes simply by recording the pressure readings which are taken at every 20 millilitres of volume introduced Such a record might read

4 6 8 10^{D} 11 13 12^{DU} 15 17 21^{IM}
Sensation normal Total volume 10 units—200 millilitres

or with unperceived systoles

2 7^{U} 12 15 20^{U} 28 35^{U} 40^{IVL}
Filling sensation normal Co-operation good
Terminal leak Volume 160 millilitres

It is advisable to standardize the graphic record and the illustrations are based upon a uniform plotting of pressure ordinate 5 times the volume abscissa (for illustrative examples *see* Chapter 3 and Nash 1949)

CASES OF SPECIAL COMPLEXITY

Case 1

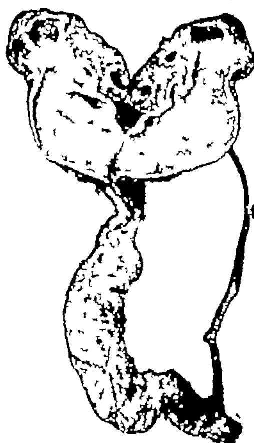
A S, girl, bladder outlet obstruction caused by intravesical protrusion of a secondary ureter

This little girl was first seen at the age of 1½ years She had had a normal gain of weight since birth and at 10 months weighed 18 lb There had been no difficulty in micturition but she was now suffering from recurrent attacks of vomiting and a craving for fluid, the urine was normal She was drinking up to 3 pints a day and on physical examination there was no abnormality beyond a prominant forehead

At the age of 2 years she weighed 20 lb and was found to have a coliform pyuria Blood urea was 58 mg per 100 ml and intravenous pyelography showed gross enlargement of the left pyelon and no excretion on the right Cystoscopy showed trabeculation and gross cystitis but the ureteric orifices were normal At this time, the girl was passing up to 14 ounces of urine at one act and although she was wet at night she was continent during the day At the age of 2½ years cystoscopy revealed that the left ureteric orifice was now dilated although the right was normal There was no residual urine Urinary infection continued unabated except for a few occasions on which the urine seemed to be clear after treatment with sulphacetamide The child's growth and weight gain were greatly retarded and by the age of 4 years the residual urine had risen to 11 ounces The diagnosis seemed to be one of idiopathic bladder-neck obstruction and an attempt was made to incise the bladder neck through the urethroscope with the use of diathermy This was unsuccessful and cystoscopy a few weeks afterwards showed 3 holes in the lower trigone resulting from the fulguration Supra-pubic cystotomy showed that these 3 holes were in the wall of an intramural cyst at the bladder neck below the trigone The roof of this cyst was removed and an accessory ureter from the right side found to be opening into it The lower opening of this cyst was evidently in the urethra but could not be found The child was now 4½ years There was no improvement following the resection of this cyst wall, which was thought to be the cause of the bladder-neck obstruction The blood urea had risen to 94 and the bladder was again opened and a wedge resected from the neck Following this procedure the residual urine was reduced to 2½ ounces but continued to be thick and ropy with mucus which was coming from the right side, and at the age of 5 years the right kidney and both its ureters were removed (*see* Fig 127 (*a*)) Ten days after this the urine became sterile and has remained sterile since

The blood inorganic phosphate was 4 1 milligrams per cent and the serum calcium 10 6 milligrams The weight at 5 years was 30 lb and height 40 inches, the specific gravity of the urine varied from 1001 to 1004 The blood urea was 72 and the residual urine was now reduced to 1 ounce Blood pressure was 110/75





(a)

(b)

FIG 127 —Case 1—A S
(a) Operation specimen showing condition of right upper urinary tract,
(b) cystogram showing reflux and vesical trabeculation.

Comment.—The first relevant symptoms in this child's history were those of polyuria and polydipsia although there was apparently no interference with the act of micturition There must have been considerable back-pressure in the urinary tract dating back to birth or even earlier Damage to the renal parenchyma must have been advanced early on and the date of the first infection is not known. Bladder-neck obstruction did not become overt until the age of 4 years and the diagnosis of ureteric ectopia was concealed by the fact that the accessory orifice, though never actually located, must have been just above the sphincteric mechanism

The damage to the urinary tract was caused by obstruction at the bladder outlet by a dilated intramural portion of an accessory ureter The child is dwarfed



FIG 128—Case 2—M S Excretion pyelogram No secretion from right side, bifid pelvis left

as a result of renal damage In spite of the gross interference with the structure of the remaining urinary tract, the urine is sterile and the child well

Case 2

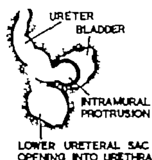
M S, boy, aged 5 years Right ectopic ureter opening in posterior urethra with intravesical protrusion causing bladder outlet obstruction

This boy had been dry by night since the age of 2 years but consistently incontinent by day Recently pyuria had developed

On admission a healthy looking little boy with day incontinence and frequency, nothing abnormal on external examination, urine infected with *B proteus*, blood urea 34 mg per 100 ml, BP 130/90 Intravenous pylogram (*see* Fig 128) showed

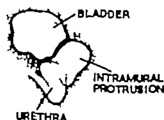


(a)



(b)

FIG 129 —Case 2—M.S.
(a) Cystogram (oblique) showing dilated ureter Intramural protrusion trabeculated bladder and large infra vesical sac
(b) explanatory drawing.



(b)

FIG 130 —Case 2—M.S
(a) Cystogram (oblique) showing bladder infra vesical sac and dilated and distorted urethra
(b) explanatory drawing



(a)



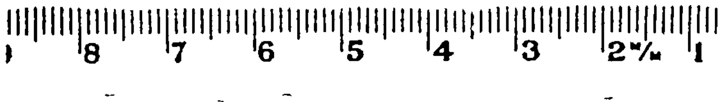


FIG 131 —Case 2—M S Aplastic right kidney Specimen removed at second operation

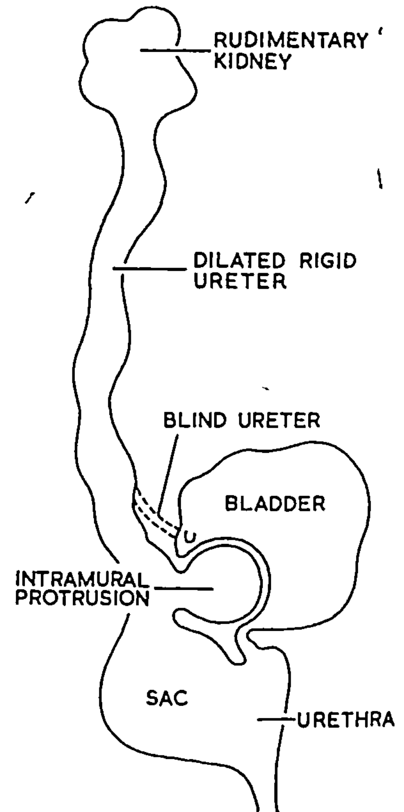


FIG 132 —Case 2—M S Key drawing

no secretion on the right side and bifid pelvis on left, with mild dilatation of the whole upper tract

Cystography (*see* Figs 129 (*a*) and 130 (*a*)) showed a small trabeculated bladder with gross dilatation of the lower end of the right ureter, an intravesical protrusion and a large sac behind and inferior to the bladder associated with marked dilatation of the posterior urethra The appearance suggested a right ectopic ureter with complex dilatation of its lower end causing some obstruction to the bladder outlet

On cystoscopy the left U O appeared normal The right half of the trigone showed a very large bulging swelling, over the top of which could be visualized a small ureteric opening drawn upwards and outwards, triangular in shape and flattened in appearance The main swelling extended down to the internal meatus, being clearly attached to the bladder wall and deforming this laterally The urethra appeared widely dilated but no orifice could be seen

Operation.—The dilated lower ureter was exposed by dissection in the pelvis and appeared to have a very broad base in the region of the bladder neck and urethra. The bladder was opened and the intravesical protrusion incised and its continuation upwards into a megaureter confirmed. Downwards the lumen ran on into the infra vesical sac which was intimately fused with the bladder wall and finally opened freely into the urethra. An attempt was made to pass a ureteric catheter into the small secondary ureteric orifice. The catheter passed easily but stuck at a distance of 2 centimetres and this orifice evidently only led into a blind ureteral stump. The megaureter was divided and the upper terminal reimplanted into the bladder. The dilated lower end was then dissected free and removed as close to the urethral wall as seemed safe and the orifice closed with cat gut stitches. The bladder was closed round a small drain and the pelvis drained.

The boy made satisfactory progress following the operation. After 8 days urine was passing per urethram and the bladder closed normally.

Following this operation the urinary infection persisted and there was still a good deal of incontinence. It was felt that the right kidney was responsible and accordingly 4 months later the boy was re admitted for nephrectomy. Rather to our surprise the kidney was found agenetic though the remnants bore every evidence of a severe suppurative infection and the ureter which was removed down to the pelvic brim was thick walled adherent and manifestly chronically inflamed (*see* Figs. 131 and 132). The urine has cleared though there is still some incontinence which is decreasing.

Case 3

S.H., girl, aged 4 months. Right pyelon duplex—ectopic ureter with orifice in urethra and large intravesical protrusion.

This infant had a severe urinary infection at the age of 6 weeks, which responded briefly to sulphamethazine but recurred and was then intractable. On admission she was pale and in poor condition with a distended abdomen. Bladder and kidneys not palpable urine infected with *B. coli* and *proteus* blood urea 38 mg. per 100 ml. Pyelography showed a right-sided duplication with hydro-nephrosis and a gross megaureter. The left kidney was normal and actively secreting. Cysto-urethroscopy showed marked cystitis with some trabeculation. The left ureteric orifice was not visualized. The right side of the bladder was occupied by a large intravesical protrusion which bulged over the internal meatus. On the apex a UO was detected. A second and much larger orifice was seen in the urethra just below the vesical neck.

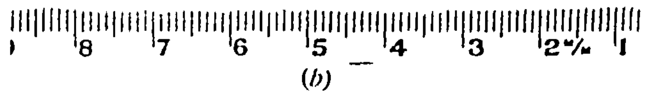
Fig. 133 (*a*) shows an attempted cystogram and is in fact a ureterogram the catheter clearly having entered the urethral ectopic orifice and the dye fills the secondary ureteral system demonstrating the cystic dilatation at the lower end.

Operation.—Right nephro-ureterectomy (specimen shown in Fig. 133 (*b*)). The ureters were divided as low down as possible no attempt being made in view of the infant's condition to complete the enucleation of the terminal ectopic segment. She made an uneventful recovery and 3 months later the urine was clear she was symptom free and in vigorous health.



(*a*)





(*b*)

FIG 133—Case 3—S H
(*a*) Retrograde ureterogram of ectopic ureter showing terminal dilatation (intravesical protrusion),
(*b*) specimen as removed at operation (gross dilatation of deuteropyelon)

Case 4

J G, boy, aged 12 years Crossed renal ectopia (calculi) associated with left hemidysplasia of upper and lower limbs

This boy, who has been under observation for several years, has a remarkable association of tragic congenital deformities

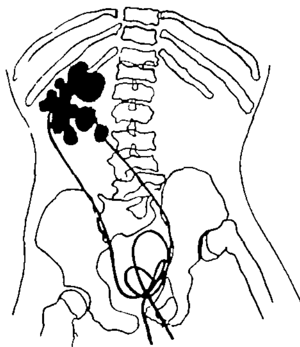
(1) Left upper limb—suppression of radius and lower ulna with radial deviation of the hand to 90 degrees

(2) Left lower limb—gross malformation of sacrum and coccyx with dislocation of the left hip and malformation of the femoral head.

(3) Crossed renal ectopia (*see* Fig. 134 (*a*) and (*b*)) both renal elements being in the right loin Both lodge calculi The boy has had much trouble from urinary infection Calculi have been removed from the bladder and from the kidney and his condition thereby improved, though the urinary infection is only partially controlled His general condition is surprisingly good despite the seriously depleted renal function



(*a*)



(*b*)

FIG 134—Case 4—J G
(*a*) Skiagram showing calculi in both segments of the crossed ectopic kidney Note also the sacro-coccygeal and femoral bony malformations
(*b*) composite pyelographic tracing showing the crossed ectopia.

Case 5

L H , boy Megaureter of doubtful nature

This boy was first admitted at 3 months of age under Dr Donald Paterson because of failure to thrive He was found to have a urinary infection (*B coli*) which cleared under sulphadiazine Blood urea at that time was 40 mg per 100 ml



(*a*)



(*b*)

FIG 135 —Case 5—L H
(*a*) Cystogram showing dilated upper tract,
(*b*) voiding cysto-urethrogram showing the same—note dilatation, particularly in the lumbar segments,

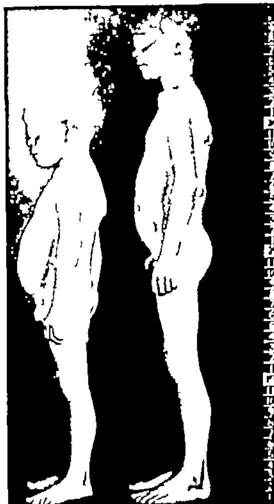
Three years later (aged 3 years and 3 months) he was re admitted with a mild attack of pneumonia when retarded growth and distended abdomen were noted Blood urea 44 mg. per 100 ml urine contained pus cells and *B coli* Intravenous pyelogram showed very poor concentration of dye A cystogram at that time partly outlined the ureters by reflux (Fig 135 (*a*)) and showed considerable dilatation in the lumbar segment One year later (aged 4 years and 3 months) polyuria cystoscopy showed a normal bladder and ureteric orifices Residual urine ½–1 ounce A voiding cysto urethrogram gave no evidence of bladder outlet obstruction but showed gross dilatation of the upper tracts (*see* Fig 135 (*b*) and (*c*)) BP normal urine sterile but contained a considerable quantity of albumin Growth continued poor and he was re-admitted at the age of 6 years dwarfed (*see* Fig. 135 (*d*)) and abdomen distended Blood urea now 144 mg per 100 ml urine clear

A few months after his discharge on this occasion he died of pneumonia and no post mortem examination was carried out

Comment —The exact pathology in this case is obscure but the story suggests severe renal parenchymal damage resulting from pyelo nephritis in early infancy The predominant dilatation of the lumbar ureteral segments suggests that the ureteric lesion resulted from infection rather than from any underlying distal obstruction



(*c*)



(*d*)

FIG 135 (contd)
(*c*) voiding cysto-urethrogram lateral view—note the normal urethra
(*d*) stature in comparison with boy of same age—note, in addition to the dwarfing, the pro tuberant abdomen

Case 6

J B , boy, aged 9 years

This child suffered from attacks of pain in the left groin from the age of 4 years At the age of 9 years investigation revealed a calculus at the lower end of the left ureter, found to be bulging from the left orifice (*see* Fig 136 (*a*)) The urine was infected and there was no evidence of bladder outlet obstruction Supra-pubic cystotomy and dilatation of the left ureteric orifice failed to remove the stone, which was later removed by an extraperitoneal approach At the age of 12 years a second stone had formed in the same site (*see* Fig 136 (*b*)) and was again removed by an extraperitoneal approach, and at the age of 13 years a third stone formed in the same place (*see* Fig 136 (*c*)) and was similarly removed by an extraperitoneal approach The function of the kidneys, which were grossly dilated, remained constant throughout Cystography showed no ureteric reflux on normal filling or tilting but gross reflux on the right side only during micturition (*see* Fig 136 (*e*) and (*f*)) Reflux on the left may have been prevented by initial damage to the left ureteric orifice either by the first extraction or by the impaction of the stone, whereas repeated flushing of the right ureter by micturition had clearly prevented stone formation on the right side At the age of 13½ years, owing to persistence of infection, the redundant ureteric loop on the left shown in Fig 136 (*d*) was removed —some 3½ inches of rigid fibrous adherent tube incapable of peristalsis At the



(*a*) (*b*)

Fig. 136 —Case 6—J B

(*a*) Initial calculus lower-end left ureter, age 9 years,

(*b*) second calculus, same position, age 12 years,

age of 14½ years after all other antiseptics had failed the urine was made sterile for the first time with chloramphenicol

The boy has remained well throughout and has grown normally The basic aetiology here is not clear but may have been bladder outlet obstruction with spontaneous cure in infancy residual effects being limited to the upper tract or the primary fault may have been at the ureteric orifices though this is unlikely since there is reflux on the right side



(c)



(d)

FIG 136 (contd)

(c) third calculus, same position, age 13 years

(d) excretion pyelogram age 13 years, showing rigid loop in left ureter which was resected

The case illustrates (a) The importance of vesico-ureteral reflux in preventing stagnation in dilated ureters when the orifices "appear" normal, (b) the relevance of rigid ureteric loops in the maintenance of infection, and (c) that the life-long presence of gross dilatation of the whole tract and persistent infection may exist without marked impairment in general health or growth

Renal function as shown by excretion pyelography is still good Urea clearance tests would be invalidated by the presence of residual urine in the large ureters



(e)



(f)

FIG 136 (contd)
(e) cystogram with no reflux
(f) cystogram during micturition showing reflux up grossly dilated right ureter

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